National Education Curriculum

Specialty Curricula

Adult Cardiac

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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.

Section I: Anatomy and Physiology of the Heart

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)
      - Anterior leaflet
        - Scallops (A1, A2, A3)
      - Posterior leaflet
        - Scallops (P1, P2, P3)
      - Papillary muscles
        - Septophobic papillary muscles
      - Chordae tendinae
      - Annulus
   ii) Tricuspid valve (TV)
      - Septal leaflet
      - Posterior leaflet
      - Anterior leaflet
      - Papillary muscles
        - Septophillic papillary muscles
      - Chordae tendinae
      - Annulus

b. Semilunar valves
   i) Aortic valve (AV)
      - Right coronary cusp (RCC)
      - Left coronary cusp (LCC)
      - Non-coronary cup (NCC)
      - Sinus of Valsalva
      - Annulus
      - Commissures
      - Subvalvular, supravalvular, LVOT
   ii) Pulmonic valve (PV)
      - Leaflets
      - Subvalvular
      - Supravalvular
      - 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 aorta
v) Aortic arch
vi) Descending aorta
vii) Abdominal aorta

d. Major branches
i) Brachiocephalic (innominate)
- Bifurcate into the right common carotid and the right subclavian artery

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. Epicardium
3. Myocardium
4. Endocardium

E. Relational Anatomy

1. Mediastinum anatomy
2. Heart position
   a. Levocardia
   b. Mesocardia
   c. Dextrocardia
   d. Levoposition
   e. Mesoposition
   f. Dextroposition
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. Ventricular looping
   a. Dextro (rightward) ventricular looping (D-loop)
   b. Levo (leftward) ventricular looping (L-loop)
4. Regions of heart tube
   a. Sinus venosus
   b. Primitive atria
   c. Atrioventricular canal
   d. Primitive LV
   e. Interventricular foramen
   f. Primitive RV
   g. Conus arteriosus
   h. Truncus arteriosus
   i. Aortic sac
   j. Aortic arches
   k. Pulmonary arteries
5. Septation
   a. Endocardial cushion
   b. Atrioventricular canal
   c. Conotruncal 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. Left Ao Arch (L) - left 4th pair

e. Proximal right subclavian (R) – partial component of the right 4th pair

f. Rudimentary versus not present in humans- 5th pair

g. Left pulmonary artery (LPA) and ductus - left 6th pair

h. Right pulmonary artery (RPA) - right 6th pair

7. Cardiac valve formation

a. The atrioventricular valves and supporting apparatus

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 2D 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) 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
         ii) Tricuspid valve (TV)
         iii) Right Atrium (RA)
         iv) RA appendage
      c. Long-axis right ventricular outflow tract (RVOT)
         i) Outlet portion of RV
         ii) Pulmonary valve
         iii) Main pulmonary artery (MPA)
      d. Short-axis
         i) High parasternal

ii) Level of aortic valve, great vessels and atria
- AV
  - Right Coronary cusp
  - Left Coronary cusp
  - Noncoronary cusp
- TV
- RVOT
- PV
- MPA bifurcation
- LA
- RA
- Interatrial septum (IAS)

iii) Coronary arteries
- RCA
- LMA
- LAD
- Circumflex artery

iv) Left atrial appendage (LAA)

v) Left ventricle
- 16 segment wall identification
- Level of the mitral valve leaflets (basal level)
  - Anterior leaflet
  - Posterior leaflet
- Papillary muscles (mid-level)
  - Anterior lateral
  - Posterior medial

vi) RV

2. Apical views

a. 4-chamber

i) Ventricles and atria

ii) MV

iii) TV

iv) Pulmonary veins

v) Coronary sinus (posterior tilt)

vi) RVOT, PV, PA (anterior tilt)

vii) IVS

viii) IAS

b. 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. Apical 4-chamber RV-focused
   i) RA
   ii) TV
   iii) RV
   iv) LA
   v) LV

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

e. Long-axis
   i) LV
   ii) LA
   iii) LVOT
   iv) MV
   v) AV

3. Subcostal views

a. 4-chamber view
   i) LV
   ii) RV
   iii) LA
   iv) RA
   v) MV
   vi) TV
   vii) IAS
   viii) 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) AV

d. Long-axis inferior vena cava (IVC)
   i) RA
   ii) IVC
   iii) Hepatic veins

4. Suprasternal views

a. Long-axis
   i) Ascending aorta
   ii) Transverse aorta
   iii) Descending aorta
   iv) Branch vessels

b. Short-axis
   i) Aortic arch
   ii) SVC
   iii) Pulmonary veins
   iv) Right pulmonary artery branch

5. Alternative views

a. Mid clavicular
b. Right parasternal
c. Posterior
d. Subcostal abdominal aorta
e. Right lateral imaging of the IVC
f. Subcostal bicaval view
g. Parasternal short-axis coronary artery view
h. SVC

C. Echocardiography Anatomy and Physiology

1. Orientation of images

a. Terminology
b. Display

2. Coronary artery distribution
3. Left ventricle (LV)

a. Dimensions, area, 2D and 3D volumes

b. LV mass, wall thickness

c. Function

i) Global systolic

- Ejection fraction
- Strain

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, 2D and 3D volumes

b. Global systolic function

i) Fractional area change

ii) Strain

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

i) Anatomic location

ii) Doppler interrogation

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 (2D) 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) Rocking
   v) Probe pressure
   vi) Sliding

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

c. Doppler tissue imaging (TDI)
i) Low wall filter setting (high pass velocity setting)
ii) Low gain settings
iii) Color flow assignment
iv) Frame rate
v) Scale adjustment
vi) Sample size or placement
vii) Respiratory variation
viii) Clinical use and formulas
ix) Waveforms
- e' - early diastolic annular velocity

- a' - annular velocity during atrial contraction
- s'- systolic annular velocity

d. Standalone dedicated CW transducer
   i) Clinical application
   ii) Recommended acquisition windows

e. Imaging planes

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

5. Three-Dimensional (3D) imaging

a. Transducer
   i) Matrix-array

b. Equipment controls
   i) Real time
   ii) Live
   iii) Simultaneous multiplane mode
   iv) Volume size
   v) Single beat, multibeat
   vi) Zoom
   vii) Full volume
   viii) ECG Gating
   ix) Cropping
   x) Rendering
   xi) 2D tomographic slices
   xii) Color flow
   xiii) Artifacts (over gain, under gain, stitch artifact, breathing)
   xiv) 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

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
    - ~ Estimation of pressure gradients
    - ~ Valvular stenosis (aortic, pulmonic)

- ~ Pulmonary artery pressure
- ~ Limitations

iii) Reynolds number

- Point at which turbulence occurs

C. Physiology and Hemodynamic Information Derived from Doppler

1. Pressure and flow resistance
2. Stroke volume (SV)
3. Cardiac output (CO)
   a. Measured in liter/minute
4. Cardiac index (CI)
   a. Measured in liters/minute/meter^2
5. dP/dt
   a. Delta pressure (dP) over delta time (dt) mmHg/msec
   b. Time in milliseconds (msec) it takes the LV to generate 32 mmHg; first derivative of LV pressure rise
6. Continuity principle
   a. Based on conservation of mass
   b. Stroke volume method
      i) Calculation of valve areas
      ii) Effective regurgitant oriface (ERO)
      iii) Regurgitant volume/regurgitant fraction
      iv) Pitfalls/limitations
7. PISA method
   a. Valve areas
   b. Effective regurgitant oriface (ERO)
   c. Regurgitant volume/regurgitant fraction
   d. Pitfalls/limitations
8. 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
9. Regurgitant fraction (RF)
   a. RF aortic valve (AV)
   b. RF mitral valve
   c. Pitfalls

10. 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
        vi) 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 contraction time (IVCT)

b. Rapid ejection (acceleration)

c. Reduced ejection (deceleration)

d. Factors that influence ventricular systole

i) Ventricular function

ii) Afterload

iii) Preload

iv) Frank-Starling's law

F. 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 2D, 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

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. Aneurysmal
5. Hyperkinesis
6. 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. Geometric assumptions
      e. Oblique measurements/angles
      f. Limited frame rate of 2D imaging
      g. LV foreshortening
      h. Translation artifact
      i. Dyssynchrony of contraction
   5. Improve endocardial delineation
      a. Harmonic imaging
         i) Understand optimizing, fundamental versus harmonic versus vendor boost imaging
      b. Ultrasound enhancing agents (UEA)

D. Assessment of Ventricular Systolic Function

1. Fractional shortening
    a. Technique
        i) M-mode
        ii) 2D
    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. 2D 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. 3D
      i) Eliminates geometric assumptions
   b. 2D Simpson's rule and modified simpson's method
      i) Biplane more accurate
      ii) Single plane
         - Suitable if LV symmetric
   c. End systolic volumes (ESV)
      i) Most reproducible volume measure
      ii) Insensitive to cardiac loading
      iii) Powerful predictor of cardiac events
      iv) Normal values
   d. End diastolic volumes (EDV)
      i) Endocardium more difficult to image at end diastole
      ii) More variable than ESV
      iii) Normal values
   e. 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) Use of UEA
8. Tissue Doppler imaging (TDI)
   a. Characteristics
   b. Appropriate technique
9. Speckle tracking echocardiography (STE)

E. LV Quantification
1. Prognosis
   a. CAD
   b. Acute MI
   c. Cardiomyopathy
      i) Volume/mass ration
2. Methods for regional function

a. 16 segment wall score
b. 17 segment wall score (perfusion)
c. Speckle tracking echocardiography (STE)

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
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
   l. Spontaneous coronary artery dissection (SCAD)
5. Therapeutic implications
   a. Guideline directed medical therapy for CHF
   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 (negative 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. 2D/3D 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) E/e'
vii) IVRT
viii) Vp using color M-mode

d. Abnormal relaxation
i) Doppler values
- AR
- AR duration < mitral a duration
- DT
- E/A
- e'
- E/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'
- E/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

i) Doppler values

- AR
- AR duration – mitral a duration
- DT
- E/A
- e'
- E/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) Parameters

ii) Limitations

iii) Normal LV filling pressures

iv) Elevated LV filling pressures

4. RV quantification

a. Dimensions and volumes

i) Basal, mid and base-apex dimensions

ii) Systolic and diastolic volumes (2D or 3D)

iii) RV wall thickness

b. Systolic function

i) Tricuspid annular plane systolic excursion (TAPSE)

ii) Tricuspid annular s' velocity

iii) Fractional area change

iv) RV strain

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

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 UEA
  - 8. Anatomy
  - 9. Normal imaging in PSAX
    - a. Left main at left posterior sinus
      - i) 4 o'clock position
    - b. RCA at right aortic sinus
      - i) 11 o'clock position SAX aortic valve level
  - 10. Normal imaging in PLAX
  - 11. 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 atherosclerosis
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

b. Diastolic dysfunction

c. 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
vi) MR
vii) Dilated LV
viii) Papillary muscle dysfunction
ix) Aneurysm
x) Pseudoaneurysm
xi) Post MI pericardial effusion (Dressler's syndrome)

xii) Ischemic cardiomyopathy

xiii) 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

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 forward cardiac output
   c. Increased LA pressure eventually leads to LA enlargement, “backwards” failure and pulmonary hypertension (PH)
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) Right heart failure
         - Hepatic congestion
         - Pulmonary edema

- RV lift

iv) 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. Rheumatic MS

i) Thickened mitral leaflets and subvalvular apparatus

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

iii) Immobility of posterior leaflet

iv) "Fish-mouth" orifice in short-axis view

v) Commissural fusion

vi) Variable degrees of valvular calcification and subvalvular thickening

b. Calcific MS

i) Calcification of mitral annulus with variable extension to leaflet and chordae

ii) Leaflet tips may appear mobile while the bases appear rigid

c. Decreased mitral diastolic opening with increased mean gradient

d. LA enlargement

e. LV small with normal function (pure MS)

f. PH

i) Secondary RA and RV enlargement

ii) Elevated TR velocity

g. Candle-flame appearance of LV inflow tract (color Doppler)

h. Coexisting valvular lesions

i. Spontaneous contrast or thrombus in LA, LAA

5. Quantitation

a. Pressure gradients (CW Doppler)

i) Peak (initial gradient)

ii) Mean

b. MV area

i) 2D or 3D 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) Iatrogenic 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

- Surgical
- Transcatheter

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 (functional)

k. Atrial remodeling (functional)

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) Late systolic murmur with MVP
        iv) S3, S4 with heart failure (in sinus rhythm)
        v) 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
            - Normally observe 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 in the absence of MVP
            - 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
      iii) 3D
         - Vena contracta area
   b. Parameters
      i) Regurgitant volume
      ii) Regurgitant fraction
      iii) Regurgitant orifice area
   c. Severity 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
- Transcatheter edge-to-edge repair (TEER)
- Alfieri stitch
- Minimally invasive

iii) MV replacement (MVR)

- Surgical
- Transcatheter

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) Many 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. Mitral annular disjunction (MAD) may be present
   d. Leaflets may be thickened or myxomatous
   e. Elongated chordae
   f. LA and/or LV enlargement with significant MR
   g. Mitral regurgitation, typically eccentric and late systolic
   h. Progression of MR severity over time
   i. Assess RVSP: indirect clue to severity of MR and compensation to volume overload
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. Medical and surgical treatment as for MR if indicated
      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 also present
        iii) S3, S4 with heart failure
4. Echocardiographic features
    a. Aortic valve 2D 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) 2D or 3D 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 flow-low gradient AS

a. Classical

i) Small calculated valve area with limited excursion of leaflets, low mean gradient differentiate factors of low stroke volume or LV systolic dysfunction

ii) Defined as an AVA of ≤1 cm^2, mean gradient <40 mmHg, Vmax of <4.0 m/s and an LVEF of <50%

iii) Two possible explanations

- True anatomically severe aortic stenosis
- Functional or pseudo severe aortic stenosis
- Possible measurement error

b. Paradoxical

i) Small calculated valve area with limited excursion of leaflets, but mean gradient is low

ii) Defined as an AVA of ≤1 cm^2 (or indexed AVA <0.6 $cm^2/m^{2)}$ mean gradient <40 mmHg, stroke volume index (SVI) of <35 mL/m^2 and an LVEF of >50%

iii) Characterize by reduced forward stroke volume

iv) Possible explanations

- Impaired LV filling
- Atrial fibrillation
- Significant mitral regurgitation
- Mitral stenosis
- Tricuspid stenosis

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. Computed tomography (CT)

i) Calcium score

e. 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) Treatment for heart failure

ii) Anticoagulation if clinically indicated

b. Surgical

i) Balloon valvuloplasty (in children)

ii) AVR

- Surgical
- Transcatheter (TAVR)

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 (in sinus rhythm)

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

d. Cardiac MRI

i) Useful if echocardiography inconclusive on AR severity

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 (surgical or transcatheter)

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

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 very large

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/repair
ii) TV replacement (surgical or transcather)

H. Pulmonary Stenosis (PS) – See Pediatric/Congenital Curriculum for Further Detail
1. Etiology
a. Most commonly congenital
b. Acquired PS
i) Carcinoid heart disease
ii) Rheumatic valve disease
2. Pathophysiology
a. Obstruction of RV emptying during ventricular systole
b. Results in increased RV pressure
3. Clinical presentation
a. Related to severity of obstruction
4. Echocardiographic features
a. Varying site of obstruction and valve morphology
b. RVH with chronic pressure overload
c. Severity evaluated by peak gradient
d. With PS or RVOT obstruction, RVSP is greater than PASP

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. Right ventricular volume overload (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
      iii) May be deceptive in severe PR due to rapid equilibration of diastolic pressure
   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

d. Cardiac MRI

i) Useful if echocardiography inconclusive on severity

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) PV replacement

J. Infective Endocarditis

1. Etiology

a. Microbial infection of the endothelial lining of the heart

b. Most common bacterial organisms

i) Streptococci

ii) Staphylococci

iii) Enterococci

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
    v) Previous endocarditis

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. 2D/3D/M-mode features
    a. Echo hallmark: vegetation
    b. Common locations
        i) Atrial (upstream) side of MV and TV
        ii) Ventricular (upstream) 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 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) Types of tissue (most common)
         - Bovine
         - Porcine
      ii) Stented

- Common examples
- Central flow
- Trivial or no regurgitation

iii) Stentless

- Intended for aortic position
- Common examples
- Very low mean gradient

iv) Transcatheter

d. Valve conduits

i) Mechanical or tissue prosthesis attached to a vessel graft or conduit

e. 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

f. Transcatheter valves

i) Size

ii) Placements

- Aortic
- Mitral
- Tricuspid

- Pulmonary
- Valve-in-valve

iii) Common examples

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. Two major categories
      i) Structural valve dysfunction
         - Intrinsic permanent changes to the prosthetic valve
         - Wear and tear
         - Leaflet fibrosis or calcification
         - Stent or strut fracture or deformation
      ii) Nonstructural valve dysfunction
         - Prosthesis-patient mismatch
         - Paravalvular leak
         - Other
   b. 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

c. 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 (physiologic) 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 (physiologic) 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
  - Mild degree often seen with TAVR

d. Thromboembolism

i) Higher risk with:
- Mechanical valves
- Atrial fibrillation
- Large LA
- LV dysfunction

e. 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

f. Endocarditis
i) Higher risk of SBE in patients with prosthetic valves
ii) Prophylaxis recommended

g. 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

h. Prosthetic structural failure
i) Poppet/cage variance
ii) Strut fracture
iii) Valve degeneration or calcification of tissue valves

i. 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. 2D/3D 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/gradient

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 aortic 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
- Using the label size of the prosthetic valve to calculate the annular cross-sectional area is not recommended

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. Doppler velocity index (DVI)

i) Index of valve performance

ii) 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

iv) Pressure recovery phenomenon

9. Doppler assessment of mitral valve 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. Doppler velocity index (DVI)

i) Index of valve performance

e. Measure pressure half-time

f. Estimate pulmonary artery pressure

g. 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. Additional imaging modalities for evaluation of prosthetic valves

    a. Cinefluoroscopy

    b. Cardiac catheterization

    c. CT

d. Cardiac magnetic resonance (CMR)

e. Cardiac positron emission tomography (PET)

14. Considerations for transcatheter and valve-in-valve prostheses types of valves

a. Echocardiography's role

b. Technical considerations

c. Differences in measurement and Doppler acquisition

d. Complications specific to transcatheter procedures

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. Etiology
   a. Genetic disorder
   b. Variable expression of disease severity
3. Pathophysiology
   a. Ventricular hypertrophy
   b. Myocardial disarray
4. Clinical presentation
   a. Signs and symptoms
   b. Physical findings
   c. Auscultatory findings
5. Types/variants
   a. Classic hypertrophic obstructive cardiomyopathy (HOCM)
   b. Midventricular
   c. Apical
      i) May demonstrate an apical “pouch”
   d. Concentric
   e. Other
6. Provocative maneuvers
   a. Standing
   b. Valsalva
   c. Amyl nitrite
7. 2D/3D/M-mode features
   a. Small or normal LV cavity
   b. Normal or hyperdynamic LV function
   c. Presence and distribution of hypertrophy
   d. Dynamic LVOT obstruction

i) Narrowed LVOT diameter
ii) Venturi effect
iii) Anterior papillary muscle displacement and elongation
iv) Systolic MV anterior motion (SAM)

e. Mitral apparatus
i) Elongated anterior mitral leaflet (AML)
ii) Calcified mitral annulus

f. LA enlargement

g. Altered texture of myocardium

h. Basal septal endocardial thickening (contact lesion)

8. Doppler features

a. Diastolic dysfunction
i) Comprehensive assessment required
ii) Characteristics
- Predominant abnormality is impaired relaxation
- Decreased compliance
- Increased LVEDP

iii) Poor correlation between DT and LA pressure
iv) 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) Mechanism: 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) Amyloid
iv) Friedreich's ataxia

b. Dynamic LVOT obstruction not specific for HCM

10. Related testing

a. ECG
b. Laboratory testing
c. Correlative imaging

11. Common treatments
a. Guideline-directed medical therapy
b. Pacemaker implementation and therapy (CRT)
c. Septal ablation (with echo guidance)
d. Surgical myectomy

B. Dilated Cardiomyopathy

1. Epidemiology
a. Prevalence
b. Genetics

2. Etiologies
a. Familial/genetic
b. Viral
c. Alcohol/toxic
d. LV non-compaction

3. Clinical presentations
a. Signs and symptoms
b. Physical findings
c. Auscultatory findings

4. 2D/3D/M-mode features
a. Left and/or right ventricular enlargement
b. Global LV systolic dysfunction
c. Eccentric LVH
d. Superimposed regional wall motion abnormalities may be present
e. Enlarged atria
f. Abnormal ventricular septal motion due to left bundle branch block
g. Possible mural thrombus in apex or aneurysm
h. Dilated mitral annulus with incomplete coaptation of the mitral valve leaflets
i. Tethering or tenting of the mitral valve leaflets due to LV remodeling
j. Spontaneous echo contrast
k. Dilated IVC with reduced inspiratory collapse

5. Doppler features
a. Decreased global systolic function indices
b. Left ventricular diastolic dysfunction
c. Associated functional mitral and tricuspid regurgitation
d. Elevated RV systolic pressure

6. Prognostic role of echo
    a. Systolic function
    b. Diastolic function
    c. RV function and RVSP
7. Related testing
    a. ECG
    b. Laboratory testing
    c. Correlative imaging
8. Common treatments

C. Restrictive Cardiomyopathy

1. Pathophysiology
    a. Abnormal diastolic function due to increased myocardial stiffness and poor compliance
2. Classifications with most common etiologies
    a. Myocardial
        i) Non-infiltrative
            - Idiopathic
        ii) Infiltrative
            - Amyloidosis
            - Sarcoidosis
        iii) Storage disease
    b. Endomyocardial
        i) Hypereosinophilic endomyocardial disease
        ii) Radiation
        iii) Anthracycline toxicity
3. Clinical presentation – consistent with biventricular heart failure
    a. Signs and symptoms
    b. Physical findings
    c. Auscultatory finding
4. 2D/3D/M-mode 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 LV and RV systolic function (early)
5. Doppler features
    a. MR and TR

b. Diastolic dysfunction
c. Typically minimal or absent respiratory variation in Doppler flows

6. Differential diagnosis
   a. Constrictive pericarditis
   b. Restrictive hemodynamics versus restrictive cardiomyopathy
7. Related testing
   a. ECG
   b. Laboratory testing
   c. Correlative imaging
8. Common treatment

D. Arrhythmogenic RV Dysplasia/Cardiomyopathy
1. Uncommon inherited disorder
2. Pathophysiology
   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 presentation
   a. Most often ventricular arrhythmias or sudden cardiac death
   b. Signs and symptoms
   c. Physical findings
   d. Auscultatory finding
4. 2D/3D/M-mode features
   a. Wide spectrum of RV involvement
   b. Dilated RV
   c. Aneurysm, outpouching of the RV
   d. Focal RV wall thinning
   e. RV wall motion abnormality
   f. RV systolic dysfunction
5. Related testing
   a. ECG
   b. Laboratory testing
   c. Correlative imaging
6. 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 of cardiac involvement
   a. Increased afterload leads to left ventricular hypertrophy (LVH) and increased mass
   b. Ventricular hypertrophy leads to diastolic dysfunction
3. 2D/3D/M-mode/color flow Doppler 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 (PH)

1. Mean pulmonary artery pressure (mPAP) > 20 mm Hg at rest

2. Classification
    a. Pulmonary arterial hypertension (PAH)
    b. Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis
    c. Pulmonary hypertension due to lung disease or hypoxia
    d. Chronic thromboembolic pulmonary hypertension
3. Pathophysiology
    a. PAH – increased pulmonary vascular resistance due to vascular remodeling or excessive vasoconstriction
    b. PH due to secondary causes
        i) Left-to-right shunts
        ii) Left heart disease
        iii) Acute pulmonary embolism
        iv) Chronic obstructive pulmonary disease (CPOD)
        v) Scleroderma
4. 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 (pressure overload)
    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
5. Eisenmenger's syndrome
    a. Reversal of intracardiac shunt from left-to-right to right-to-left secondary to severe pulmonary hypertension
6. Cor pulmonale
    a. Acute (pulmonary embolism)
    b. Chronic
7. Prognostic value of echocardiography
8. 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

X. PERICARDIAL DISEASES

- A. Normal Pericardium
  - 1. Structure
  - 2. Function
- B. Pericarditis/Pericardial Effusion
  - 1. Etiologies
  - 2. Clinical presentation
    - 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. Pathophysiology/hemodynamics

a. Accumulation of pericardial fluid causes pericardial pressure to rise and compromise systemic venous return in right atrium

b. Intrapericardial pressure is influenced by both volume of fluid and rate at which it accumulates

c. Ventricular filling is impaired and cardiac output is decreased

d. Dissociation of intrathoracic and intracardiac pressures due to insulating effect of effusion

e. Exaggerated ventricular interdependence

4. 2D/3D/M-mode echocardiographic features

a. Pericardial effusion

b. RA or RV 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 due to ventricular interdependence

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 presentation
   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. 2D/3D/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
   i. Reduced inferior and lateral strain due to pericardial tethering
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 due to decreased radial expansion of the heart

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
b. Medical

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

XI. CARDIAC TUMORS/MASES

- 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. Use of ultrasound enhancing agents (UEA)
  - 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. DISEASE 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. 2D 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. Regional wall motion abnormality (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. Intramural 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. Explain how the segmental approach is utilized to identify and characterize congenital heart disease
2. List the standard views included in a pediatric echocardiogram
3. Describe echocardiographic findings associated with various types of congenital heart disease
4. Discuss clinical signs and symptoms associated with congenital heart disease
5. List the parameters used in qualitative and quantitative assessment of congenital heart disease for each type described in this outline

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 "indicator mark" refers to the orientation ridge on the transducer. Indicator mark is used with a clock reference for each view
   B. The term "vertex up and vertex down" refers to the ultrasound image on the display monitor. When imaging congenital heart disease in the pediatric echocardiography laboratory, images are usually orientated in the "anatomical correct" position on the display monitor.
3. When describing "sweeping" or "angle", 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 inferior vena cava, superior vena cava, and coronary sinus
      i) Pectinate muscles within chamber
      ii) Right atrial appendage is broad-based, triangular shape
2. Left atrial morphology
   a. Receives pulmonary veins
   b. Smooth walled; pectinate muscles limited to the left atrial appendage
   c. Left atrial appendage is thin, narrow-based shape, pectinate muscles within chamber
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 tricuspid and pulmonic valve

5. Left ventricular morphology
    a. Smooth superior septal surface
    b. Fine endocardial trabeculations
    c. Fibrous continuity between mitral valve and aortic valve
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 {S, D, S} – first letter of segmental set
    a. S - situs solitus; morphologic right atrium is right-sided, morphologic left atrium is left-sided
    b. I - situs inversus; morphologic right atrium is left-sided, morphologic left atrium is right-sided
    c. A - situs ambiguous; associated with isomerism of the atrial appendages
2. Ventricular segment {D, L, X} – second letter of segmental set
    a. D – loop; morphologic right ventricle on the right side
    b. L – loop; morphologic left ventricle on the right side
    c. X – unable to determine looping
3. Great artery segment {S, I, D, L, A} – third letter of segmental set
    a. S – solitus; normal position of the aortic valve, rightward and posterior to the pulmonic valve
    b. I – inversus; aortic valve is leftward and posterior to the pulmonic valve
    c. D – d-malposed; aortic valve is rightward and anterior to the pulmonic valve
    d. L – l-malposed; aortic valve is leftward and anterior to the pulmonic valve
    e. A – anterior; aortic valve directly anterior to the pulmonic valve

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 concordance verses discordance

b. Ventriculoarterial concordance verses discordance

c. Atrioventricular and ventriculoarterial discordance (congenitally “corrected” transposition of the great vessels)

5. Assessment of cardiac structural morphology

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

b. Display all structures in correct anatomical position

D. Imaging Technique

1. Subcostal

a. Coronal View - situs view (vertex up or down); transverse cut

b. Indicator marker at 3 o’clock

i) 2D + color clip: cross-section of inferior vena cava, descending aorta, spine, liver, stomach

ii) 2D + color sweep: hepatic veins to inferior vena cava to right atrium; continue to sweep to demonstrate cardiac apex

c. Sagittal view - inferior vena cava and descending aorta view (vertex up or down)

d. Indicator marker 12 o’clock (vertex up); 6 o’clock (vertex down)

i) 2D + color clip: inferior vena cava

ii) 2D + color clip: abdominal descending aorta

iii) PW: abdominal descending aorta below diaphragm

e. Coronal view- long-axis view (vertex down)

f. Indicator marker 3 o’clock; posterior to anterior sweep

i) 2D + color sweep: inferior vena cava to right ventricular outflow tract

ii) Focused 2D + color sweep at low nyquist limit: atrial septum

g. Left anterior oblique view

h. Indicator marker between 4-5 o’clock; posterior to anterior sweep

i) 2D + color sweep: atrial septum

ii) 2D + color clip: tricuspid valve and mitral valve en face

i. Sagittal View- short-axis view (vertex down)

j. Indicator maker 6 o’clock, right to left sweep

i) 2D + color sweep: bicaval view to left ventricle apex

ii) Focused 2D + color sweep at low nyquist limit: atrial septum

iii) 2D + color clip: right upper pulmonary vein

iv) 2D + color clip (+/- PW, CW): right ventricular outflow tract

k. Right anterior oblique view

l. Indicator marker between 1-2 o’clock

i) 2D + color clip: right ventricle inflow and outflow

2. Apical views (vertex down)
    a. 4-chamber
    b. Indicator marker 3 o'clock; posterior to anterior sweep
        i) 2D + color sweep: coronary sinus to right ventricular outflow tract
        ii) 2D + color sweep at low nyquist limit: ventricular septum
        iii) Focused 2D clip: right atrium and right ventricle (right ventricle-centered view)
        iv) 2D + color clip: tricuspid valve
        v) PW + CW (+/-) tissue Doppler: tricuspid valve
        vi) CW: tricuspid regurgitation jet (if present)
        vii) 2D + color clip (+/-PW, CW): right ventricular outflow tract
        viii) Focused 2D clip: left atrium and left ventricle
        ix) Color + PW: one right pulmonary vein
        x) 2D + color clip: mitral valve
        xi) PW + CW + tissue Doppler: mitral valve
        xii) Measurements: tricuspid and mitral valve annular diameters; left atrial end-systolic area; left ventricular end-diastolic and end-systolic area and length, left ventricular end-diastolic epicardial length; right ventricular end-diastolic and end-systolic area, tricuspid annular plane systolic excursion
    c. 2- chamber
    d. Indicator marker 2 o'clock
        i) 2D clip: left atrium and left ventricle
        ii) 2D + color clip: mitral valve
        iii) Measurements: left atrial end-systolic area; left ventricular end-diastolic and end-systolic area and length
    e. 3- chamber
    f. Indicator marker 11 o'clock
        i) 2D + color clip: left ventricular outflow tract, aortic valve, and proximal aorta PW + CW: left ventricular outflow tract, aortic valve, and proximal aorta
3. Parasternal views (vertex up)
    a. Parasternal long axis
    b. Indicator marker 10 o'clock; sweep inferiorly towards right ventricular inflow, then sweep anteriorly towards right ventricular outflow tract
        i) 2D + color sweep: reference to tricuspid valve, back to reference to pulmonic valve
        ii) Focused color sweep: ventricular septum
        iii) 2D + color clip: mitral valve
        iv) 2D + color clip: aortic valve
        v) 2D + color clip: tricuspid valve
        vi) CW: tricuspid regurgitation jet if present
        vii) 2D + color clip: pulmonic valve

viii) PW + CW: PV

ix) Measurements: aortic valve annular, aortic root, sinotubular junction, and ascending aorta diameters; mitral and tricuspid valve annular diameters; pulmonic valve annular and main pulmonary artery diameters

c. Parasternal short axis

d. Indicator marker at 2 o'clock; sweep from cardiac base to apex

i) 2D + color sweep: aortic valve to apex then back to aortic valve to main and branch pulmonary arteries

ii) Focused color sweep: ventricular septum

iii) 2D + color clip: aortic valve

iv) Focused simultaneous 2D + color clip (color-compare): left main, left anterior descending, and circumflex coronary arteries

v) Focused simultaneous 2D + color clip (color-compare): right coronary artery

vi) 2D + color clip: mitral valve

vii) 2D + color clip: tricuspid valve

viii) CW: tricuspid regurgitation if present

ix) 2D + color clip: pulmonic valve

x) PW + CW: pulmonic valve, main pulmonary artery, proximal branch pulmonary arteries

xi) 2D clip and M-mode: LV cross-section at level of papillary muscles

xii) Measurements: left ventricular end-diastolic and end-systolic areas, left ventricular end-diastolic epicardial area; left ventricular M-mode end-diastolic and end-systolic internal diameters, left ventricular M-mode end-diastolic septal and posterior wall thickness; pulmonary valve annular, main pulmonary artery, and proximal branch pulmonary artery diameters; left main, proximal left anterior descending, and proximal right coronary artery diameters

e. High left parasternal sagittal (ductal) view (vertex up)

f. Indicator marker at 12 o'clock

i) 2D + color sweep: ascending aorta to main pulmonary artery to left pulmonary artery and aortic arch to patent ductus arteriosus (if present) to descending aorta

ii) PW + CW: patent ductus arteriosus if present

4. Suprasternal notch views

a. Suprasternal short axis (vertex up)

b. Indicator marker at 3 o'clock; sweep superiorly from right pulmonary artery to brachiocephalic arteries

i) 2D + color sweep: ascending aorta superiorly to first branch bifurcation, showing arch descending to the right or left during sweep with color mapping

ii) 2D + color sweep: leftward aspect of left innominate vein (exclude left superior vena cava or partial anomalous pulmonary venous return to left innominate vein)

iii) Focused 2D + color clip (+/-) PW: two right and two left pulmonary veins draining into the left atrium (crab view; caution: right middle pulmonary vein may be confused with right upper pulmonary vein)

c. Suprasternal long axis (vertex up)

d. Indicator marker at 12 o'clock; sweep leftward towards left pulmonary artery

i) 2D + color clip: aortic arch and its branches

ii) PW + CW: distal transverse aortic arch, aortic isthmus, and proximal descending aorta

iii) Measurements: proximal and distal transverse aortic arch and aortic isthmus diameters

e. Right parasternal sagittal view (vertex up)

f. Indicator marker at 12 o'clock; right side of sternum

i) 2D + color clip: superior vena cava, inferior vena cava, atrial septum (exclude sinus venosus defect)

g. Right parasternal transverse view (vertex up)

h. Indicator marker at 3 o'clock; right side of sternum

i) 2D + color clip: right upper pulmonary vein draining into left atrium below level of right pulmonary artery

5. Calculations

a. Calculations

b. Left ventricular end-diastolic and end-systolic endocardial volumes

c. Left ventricular end-diastolic epicardial volume

d. Left ventricular mass

e. Left ventricular mass-to-volume ratio

f. Left ventricular ejection fraction (5/6 area-length method or method of disks)

g. Left ventricular m-mode shortening fraction

h. Left atrial volume

i. Right ventricular systolic pressure from tricuspid regurgitation gradient or from ventricular septal defect gradient

j. Pulmonary artery systolic pressure from patent ductus arteriosus gradient

E. Z-scores

1. Most parameters of cardiovascular size, function, and blood flow patterns change with total body growth and maturation in children

2. Z-scores for echocardiographic measurements are available for children as they grow

3. A z-score of +2 or -2 represents a measurement value that is two standard deviations above or below the mean value, usually recognized as the thresholds for normal in a pediatric population.

4. Echocardiographic z-scores should be used in children, for trending measurements over time in the same patient and when assessing risk in a particular patient population

F. Congenital Heart Disease

1. Anomalies of venous return

a. Anomalies of systemic venous return

b. Left superior vena cava draining into right atrium via coronary sinus

c. Left superior vena cava draining into left atrium

d. Retroaortic innominate vein

e. Levoatrial cardinal vein

f. Right superior vena cava to left atrium

g. Superior vena cava aneurysm

h. Interruption of the inferior vena cava

i. Absence of hepatic segment with azygous vein continuation

j. Absence of the abdominal segment with hemiazygous continuation

k. Inferior vena cava draining into left atrium

l. Absence of right superior vena cava

m. Absence of hepatic segment of inferior vena cava blood flows from inferior vena cava to azygous vein to left superior vena cava to left atrium

n. Hepatic veins drain into left atrium

o. Common atrium

2. Etiology

a. Prevalence

i) Isolated anomalies occur in 0.3-0.5% of general population

ii) Occurs in 2-10% of patients with other forms of congenital heart disease

iii) Occurs in >70% of patients with heterotaxy syndrome

b. Abnormalities in regression of umbilical, vitelline, or cardinal venous segments during embryologic development

c. Abnormalities of visceroatrial development; bilateral “sidedness” (heterotaxy) syndrome

3. Pathophysiology

a. Normal physiology of systemic venous return to the right atrium

i) Left superior vena cava to an intact coronary sinus with intact right superior vena cava

ii) Left superior vena cava to an intact coronary sinus with atresia of the right superior vena cava

iii) Interrupted inferior vena cava with azygous continuation

iv) Absent right superior vena cava with left superior vena cava to coronary sinus

v) Abnormal physiology with systemic and pulmonary venous mixing

vi) Left superior vena cava draining into left atrium

vii) Right superior vena cava draining into left atrium

viii) Left superior vena cava to an “unroofed” coronary sinus

ix) Inferior vena cava draining into left atrium

4. Clinical presentation

a. Left superior vena cava to the coronary sinus or interrupted inferior vena cava usually present with additional structural cardiac defects; can be incidental findings

b. Cyanosis is present with left/right superior vena cava or inferior vena cava draining directly to left atrium

5. Associated lesions
    a. Coarctation of the aorta
    b. Mitral valve abnormalities, including congenital mitral stenosis
    c. Heterotaxy syndrome with polysplenia
    d. Hypoplastic left heart syndrome
6. Echocardiographic evaluation
    a. Subcostal coronal (transverse) and sagittal view of the abdominal viscera to image hepatic veins, inferior vena cava
    b. Subcostal sagittal view "caval" view to assess superior and inferior vena cava
    c. Suprasternal and high right/left parasternal views (vertex up) to image superior venous anatomy
        i) Dilation of the coronary sinus in subcostal coronal, apical 4-chamber, and parasternal long views may indicate drainage of left superior vena cava into the coronary sinus
7. Treatment (surgical repair)
    i) No repair necessary if systemic venous return is physiologically normal
    ii) Intra-atrial baffle repair of left/right superior vena cava or inferior vena cava to left atrium
    iii) Ligation of left superior vena cava draining to left atrium if bilateral superior vena cava with adequate "bridging" innominate vein
8. Echocardiographic assessment postoperatively
    a. Evaluation of patency of repaired venous structures
    b. Evaluation for baffle leaks with baffle procedures
    c. Assessment of ligated venous structures

G. Anomalies of Pulmonary Venous Return

1. Types
    a. Normal pulmonary venous connections with abnormal drainage
        i) Extreme posterior (leftward) malinsertion of the atrial septum primum
    b. Partial anomalous pulmonary venous return (PAPVR)
        i) Drainage of one or more pulmonary veins anomalously to the systemic venous system or right atrium with normal drainage of remaining veins to the left atrium
        ii) Scimitar syndrome - right pulmonary veins drain infracardiac to the hepatic system or inferior vena cava (IVC)
    c. Total anomalous pulmonary venous return (TAPVR)
        i) Supracardiac
            - Pulmonary venous drainage into a vertical vein, coursing superiorly to innominate vein, draining to the superior vena cava
        ii) Cardiac
            - Pulmonary venous drainage to the posterior aspect of the right atrium
            - Pulmonary venous drainage into the coronary sinus

iii) Infradiaphragmatic

- Pulmonary venous drainage into a vertical vein, coursing inferiorly to portal venous system, ductus venosus, hepatic veins, or inferior vena cava (IVC)

d. Mixed type

i) Combination of anomalous sites of pulmonary venous return

2. Etiology

a. Incidence

i) Partial anomalous pulmonary venous return (PAPVR): up to 25% of population may have a single right or left upper vein draining anomalously

ii) Total anomalous pulmonary venous return (TAPVR): 9 in 100,000 of population

b. Occurs mostly sporadically

c. Failure of embryologic incorporation of one or more pulmonary veins to the LA

d. Partial anomalous pulmonary venous return (PAPVR) can be associated with Turner and Noonan syndromes

e. Total anomalous pulmonary venous return (TAPVR) can be associated with allagille, Trisomy 22 (cat-eye), Holt-Oram, and polysplenia/asplenia syndromes

3. Pathophysiology

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

b. Pulmonary overcirculation and congestion

c. Pulmonary hypertension (PHTN)

4. Clinical presentation (pediatric and adult patients)

a. Tachypnea/dyspnea

b. Cyanosis

c. Congestive heart failure associated with supracardiac total anomalous pulmonary venous return (TAPVR)

d. “Snowman” sign on x-ray (supracardiac total anomalous pulmonary venous return (TAPVR))

e. “Scimitar” sign on x-ray (Scimitar syndrome)

5. Associated lesions

a. Hypoplastic left heart syndrome

b. Heterotaxy syndrome with asplenia

c. Coarctation of the aorta

6. Echocardiographic assessment (pediatric and adult patients)

a. Subcostal coronal and sagittal views (vertex down) to assess confluence posterior to left atrium, atrial septum, superior and inferior vena cava

b. Suprasternal short-axis view (vertex up) to assess leftward aspect of left innominate vein for partial anomalous pulmonary venous return (PAPVR) or total anomalous pulmonary venous return (TAPVR) to left innominate vein

c. Suprasternal short-axis view (vertex up) crab view to assess individual pulmonary veins

d. Echocardiographic findings may include dilated innominate vein, superior vena cava, coronary sinus, inferior vena cava, right atrium, right ventricle, and main pulmonary artery

e. Partial anomalous pulmonary venous return (PAPVR)- define pulmonary venous return; trace anomalous pulmonary vein(s) to their point of termination

f. Total anomalous pulmonary venous return (TAPVR) - assess confluence of pulmonary veins posterior to the LA

g. Identify individual veins and their course

h. Evaluate for vertical vein - venous venophasic flow going away from the heart (superiorly or inferiorly)

i) Evaluate for obstruction along anomalous drainage pathway using color and spectral Doppler

i. Treatment (surgical repair)

i) Supracardiac and infradiaphragmatic type total anomalous pulmonary venous return (TAPVR) - direct anastomosis of pulmonary vein confluence to left atrium

ii) Cardiac (coronary sinus) type total anomalous pulmonary venous return (TAPVR) - unroofing of coronary sinus with closure of interatrial communication

iii) Scimitar syndrome partial anomalous pulmonary venous return (PAPVR) - intra atrial baffle of right pulmonary veins to left atrium

7. Echocardiographic assessment postoperatively (pediatric and adult patients)

a. Evaluate for flow obstruction at anastomosis site of pulmonary venous confluence to left atrium, pulmonary vein orifices and individual pulmonary veins

b. Evaluate for residual vertical vein

c. Evaluate for pulmonary venous baffle obstruction

d. TEE may be indicated for diagnostic quality imaging in adult patients

H. Anomalies of the Atria

1. Atrial septal defect (ASD)

a. Types

i) Patent foramen ovale (PFO)

ii) Secundum atrial septal defect (ASD)

iii) Primum atrial septal defect (ASD)

iv) Sinus venosus defect (SVD)

v) Coronary sinus defect

b. Etiology

i) Defects of the atrial septum occur in 1:1000 of population

ii) PFO found in 25% of population

iii) Primum and secundum type atrial septal defects make up 67% of all ASDs

c. Patent foramen ovale (PFO)

i) Overlapping of septum primum and septum secundum

ii) Located in the fossa ovalis region

d. Secundum atrial septal defect (ASD); mostly sporadic causes

i) Most common form of atrial septal defect

ii) Defect within the fossa ovalis associated with deficient septum primum

iii) Usually due to single or multiple defects in septum primum

iv) Septum secundum usually well-formed

e. Primum atrial septal defect (ASD)

i) Failure of septum primum to fuse with endocardial septation of the atrioventricular canal above the level of atrioventricular valves

ii) Located between the inferior septum primum and atrioventricular valves, atrial septum

f. Sinus venosus defect

g. Superior vena cava (SVC) type sinus venosus defect

i) Approximately 8% of all types of ASDs

ii) Sinus venosus not properly incorporated into the Right atrium

iii) Deficient wall between the superior vena cava and right upper pulmonary vein

iv) Superior vena cava (SVC) overrides right and left atrium

v) Located in superior and posterior portion of atrial septum

vi) Associated with partial anomalous pulmonary venous return (PAPVR)

h. Inferior vena cava (IVC) type sinus venosus defect

i) Occurs less frequently than superior type

ii) Absent sinus venosus septum

iii) Anomalous drainage of right pulmonary veins to right atrium

i. Coronary sinus (CS) defect

i) Rare

ii) Partial or complete unroofing of the tissue separating the coronary sinus from the left atrium

iii) Located between the inferior limbic septum, pars atrioventricular is of membranous septum and inferior vena cava (IVC) orifice

iv) Coronary sinus defect (CSD) and persistent left superior vena cava (LSVC) is termed Raghib syndrome

j. 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 atrial septal defects)

k. Conditions that increase left to right shunting through atrial septal defect

i) Left ventricular hypertrophy

ii) Cardiomyopathy

iii) Mitral valve stenosis
iv) Mitral valve regurgitation
v) Mitral valve atresia
vi) Cor-triatriatum sinister
vii) Aortic valve stenosis
viii) Coarctation of the aorta
ix) Volume overload
x) Pressure overload

l. Clinical presentation
i) May be asymptomatic for many years
ii) Shortness of breath (SOB) on exertion
iii) Recurrent respiratory infections
iv) Atrial arrhythmia (especially atrial fibrillation)
v) Systolic murmur secondary to increased flow across the pulmonary valve
vi) Large shunts may have diastolic murmur through tricuspid valve (TV)
vii) May have wide fixed-split second heart sound

m. Associated lesions
i) Secundum atrial septal defect (ASD) and patent foramen ovale (PFO)
- Mitral valve prolapse (MVP)
- Tricuspid atresia
- Hypoplastic left heart syndrome
- Total anomalous pulmonary venous return
- Ebstein's anomaly

ii) Primum atrial septal defect (ASD)
- Atrioventricular septal defect
- Cleft mitral valve
- Atrioventricular valve (AVV) regurgitation

iii) Sinus venosus defect
- Anomalous pulmonary venous return

n. Echocardiographic evaluation (pediatric and adult patients)
i) Subcostal coronal, left anterior oblique, sagittal views (vertex down) to assess coronary sinus, atrial septum, superior and inferior vena cava
- Deficiency in atrial septal tissue
  - Type of defect
  - Size of defect
  - Deficiencies in remaining atrial septal tissue "rims"
- "Overriding" superior vena cava in setting of superior type sinus venosus defect

ii) Apical 4-chamber view (vertex down) to assess coronary sinus, septum primum defects

- Atrioventricular valve morphology and function in relation to atrial septal defect

iii) Parasternal short axis (vertex down) to assess atrial septum

- Retro-aortic tissue "rim"
- M-mode
  - Enlarged RV
  - Paradoxical septal motion
- Evaluate for volume overload
  - Evaluate for diastolic ventricular septal flattening
- Evaluate for pressure overload
  - Evaluate for systolic ventricular septal flattening

iv) Suprasternal and high right/left parasternal views (vertex up) to image superior venous anatomy, atrial septum

- Evaluate for systemic and pulmonary venous anomalies
- "Overriding" superior vena cava in setting of superior type sinus venosus defect

o. Echocardiographic findings may include dilated right atrium, right ventricle, and main pulmonary artery; tricuspid and pulmonic valve annulus (over time)

p. May need agitated saline (microcavitation) imaging

i) Useful to detect transient right to left shunt through atrial septal defect (ASD) and patent foramen ovale (PFO)

ii) Useful to evaluate for negative contrast in the RA with left to right shunt

q. Transesophageal echocardiography (TEE) may be indicated

i) Evaluate type of defect

ii) Monitor intraprocedural progress and guidance for device closure

r. Treatment (pediatric and adult patients)

i) Catheterization

- Device closure (select cases of secundum atrial septal defects)

ii) Surgical

- Primary (suture) closure
  - Smaller defects
- Patch repair
- Baffle-type repair

s. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for residual shunt in multiple views

ii) Evaluate for atrioventricular valve regurgitation

iii) If device closure, evaluate for device impingement on surrounding structures in multiple views

iv) If sinus venosus defect repair, evaluate for superior vena cava 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" left atrium) 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 atrial septal defect (ASD) is present, the chamber may not be dilated, as the atrial septal defect 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) Atrial septal defect (ASD)

ii) Left superior vena cava (LSVC) to the coronary sinus

iii) Hypoplastic left heart syndrome (HLHS)

iv) Shone's complex

f. Echocardiographic evaluation (pediatric and adult patients)

i) Subcostal coronal, left anterior oblique, sagittal views (vertex down) to assess atrial septal defect (if present), membrane

ii) Apical 4-chamber view (vertex down) to assess membrane

- Doppler interrogation of fenestration gradient (if present)

iii) Parasternal long axis (vertex up) to assess membrane presence and point of pulmonary venous channel egress

- May stretch obliquely bisecting left atrium into pulmonary venous (dorsal) and "true" LA chambers

iv) Echocardiographic findings may include dilated right atrium, right ventricle, assess for signs of pulmonary hypertension

v) Treatment

- No treatment necessary if membrane non-obstructive

- Surgical resection of obstructive membrane with atrial septal defect closure

vi) Echocardiographic assessment postoperatively (pediatric and adult patients)

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

I. Atrioventricular Valve Anomalies

1. Atrioventricular septal defect (AVSD)

a. Etiology

i) Abnormal development of endocardial cushions

ii) 0.19:1000 live births

iii) 4.0-5.0% of all congenital heart defects (CHD)

iv) 40% of children with trisomy 21 and congenital heart disease have an atrioventricular septal defect

v) Failure of septum primum to fuse with endocardial cushions

b. Categorized as complete, partial (incomplete), or transitional (intermediate)

i) Complete atrioventricular septal defect (AVSD) consists of primum atrial septal defect, common atrioventricular valve, inlet ventricular septal defect

- Balanced
- Unbalanced
  - Left ventricular dominant
  - Right ventricular dominant

ii) Partial (incomplete) type atrioventricular septal defect (AVSD) consists of primum atrial septal defect, common atrium, cleft mitral valve

iii) Transitional (intermediate) type atrioventricular septal defect (AVSD) consists of two complete atrioventricular valve orifices, primum atrial septal defect, restrictive ventricular septal defect

c. Pathophysiology

i) Defect between the two atria and/or ventricles based on type of atrioventricular septal defect

ii) Incomplete type same as atrial septal defect (ASD), complete type same as ventricular septal defect (VSD)

iii) Volume overload

iv) Pressure overload

d. Clinical presentation (pediatric and adult patients)

i) Signs and symptoms

- Partial (incomplete) type similar to atrial septal defect (ASD)
- Complete type same as ventricular septal defect (VSD)
- Small/restrictive shunts may be asymptomatic

e. Echocardiographic evaluation (pediatric and adult patients)

i) Subcostal coronal, sagittal views (vertex down) to assess atrial septal defect(s), atrioventricular valve(s), ventricular septal defect(s)

ii) Subcostal left anterior oblique view (vertex down) to assess enface view of common atrioventricular valve

- Allows visualization of leaflet components, chordal attachments, and atrioventricular valvular commitment to each ventricle
- Images to perform atrioventricular valve index (AVVI)

iii) Apical 4-chamber view (vertex down) to assess atrioventricular valve commitment/attachments to the ventricles, presence of left ventricular outflow tract obstruction (LVOTO)

- Defect located at the cardiac crux

iv) Parasternal long axis (vertex up) to assess left ventricular outflow tract, and chordal attachments

- Left ventricular outflow length is elongated because of the unwedged aortic valve associated with a complete atrioventricular septal defect
- Assess for parachute-like attachments

v) Parasternal short axis (vertex up) to assess atrioventricular valve morphology, papilary muscle size and location

- Assess for cleft, double orifice

f. Echocardiographic findings should define:

i) Type of defect (complete, transitional/intermediate, partial/incomplete)

ii) Size of ventricles (balanced or unbalanced)

iii) Degree of atrioventricular valve regurgitation

b. Assess for signs of pulmonary hypertension

i) Ventricular septal defect gradient

ii) Right atrioventricular valve regurgitation jet

c. Related testing

i) ECG

- Counterclockwise frontal plane
- Left axis deviation
- Ventricular hypertrophy
  - Dependent upon volume of left-right shunt
  - Degree of pulmonary hypertension
  - Degree of atrioventricular valve regurgitation

ii) Chest x-ray

- Small atrial septal defect (ASD)-normal
- Large shunt – enlarged heart, increased pulmonary markings

iii) Cardiac catheterization

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

d. Treatment (pediatric and adult patients)

i) Surgical patch repair of atrial septal defect(s) (ASD) (and ventricular septal defect (VSD), if present)

- Single-patch technique

ii) Suture repair of "cleft" in anterior leaflet of left atrioventricular valve

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

iv) Resection or relief of subaortic obstruction, if present

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

i) Evaluate for residual shunt

ii) Evaluate for atrioventricular valve stenosis and regurgitation

iii) Evaluate for signs of pulmonary hypertension

iv) Evaluate for aortic valve regurgitation

v) Evaluate for left ventricular outflow tract obstruction (LVOTO)

vi) Evaluate ventricular function

2. 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 atrial septal defect (ASD)
  - Cyanosis may be present if shunt across atrial septal defect (ASD) is right to left

b. Pathophysiology

i) Varying degrees of tricuspid regurgitation

ii) Systemic venous congestion with hepatomegaly

c. Clinical presentation (adult and pediatric patients)

i) Mild forms may be asymptomatic (present in adolescence or adulthood)

ii) Severe forms (presenting as a fetus or neonate)

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

iii) Split first and second heart sound

iv) Arrhythmias - supraventricular tachycardia (SVT)

d. Associated lesions

i) Patent foramen ovale (PFO) or secundum atrial septal defect (ASD), 30-78%

ii) Pulmonary valve (PV) stenosis

iii) Pulmonary valve atresia or "functional" atresia

e. Echocardiographic evaluation (pediatric and adult patients)

i) Subcostal coronal, sagittal views (vertex down) to assess atrial septal defect(s), septal leaflet of tricuspid valve displacement and attachments, extent of atrialized right ventricle, ventricular septal defect(s)

ii) Subcostal left anterior oblique view (vertex down) to assess enface view of tricuspid valve

- Allows visualization of leaflet components, and chordal attachments

iii) Subcostal right anterior oblique view (vertex down)

- Allows visualization of the tricuspid valve, chordal attachments into the right ventricular outflow tract (RVOT), and pulmonic valve

iv) Apical 4-chamber view (vertex down) to assess right atrium and right ventricle, determine portion of "atrialized" right ventricle (RV), visualize the degree of tricuspid valve, apical displacement

- Perform Celermajer index

v) Parasternal long and short axis (vertex up) to assess right atrium, right ventricle, and tricuspid valve, right ventricular outflow tract, and pulmonary valve

- Evaluate tricuspid valve regurgitation
- Demonstrate flow (or lack of) across pulmonic valve

vi) Echocardiographic findings may include paradoxical ventricular septal motion, large right atrium (depending on the severity of apical displacement), assess for signs of pulmonary hypertension

f. Related testing

i) ECG

- Tall P waves in limb lead II
- Right bundle branch block (RBBB)
- 30% will have Wolf-Parkinson-White (WPW) syndrome

ii) Chest x-ray

- Cardiomegaly with prominent right sided border
- Decreased pulmonary blood flow

g. Treatment

i) No treatment necessary in mild cases

ii) Severe cases, an initial palliation, Blalock-Thomas-Taussig shunt (BTTS)

iii) Surgical repair (cone, Danielson, Carpentier procedures)

iv) Valve repair/replacement

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for residual tricuspid valve (TV) insufficiency and/or iatrogenic tricuspid valve (TV) stenosis

ii) Evaluate for residual atrial/ventricular level shunt, if history of atrial septal defect (ASD)/ventricular septal defect (VSD) repair

iii) Evaluate right ventricular outflow tract, pulmonic valve, and branch pulmonary arteries

iv) Evaluate ventricular function

v) Evaluate for signs of pulmonary hypertension

3. Tricuspid valve atresia

a. Etiology

i) Characterized by absence of the tricuspid valve resulting in a lack of direct communication between the right atrium and right ventricle with hypoplasia of the right ventricle

ii) 0.039-0.085:1000 live births 2.7% of all congenital heart disease (CHD)

iii) Initial survival is dependent upon interatrial communication

iv) May occur with normally related great vessels or transposition of great vessels

v) May occur with a normal pulmonary artery and large ventricular septal defect (VSD) or pulmonary stenosis and small ventricular septal defect (VSD) or pulmonary atresia. by adulthood will have gone through surgical palliation(s)

- Fontan procedure

b. Pathophysiology

i) Atresia of the tricuspid valve systemic venous return travels though atrial communication - patent foramen ovale (PFO) or atrial septal defect (ASD) to left atrium

ii) Desaturation due to systemic venous return mixing with pulmonary venous return to left atrium

c. Ventriculo-arterial relationships and the size of the ventricular septal defect (VSD) will largely determine clinical presentation

i) Restrictive ventricular septal defect (VSD) will result in subaortic stenosis in d-transposition of the great arteries

d. Ventricular volume overloading; dependent on amount of pulmonary flood flow

i) Ventricular enlargement, increases ventricular wall stress, which results in compensatory hypertrophy

- Hypertrophy – impaired diastolic function
- Subendocardial ischemia

ii) Transposed great arteries

- Aorta may arise from hypoplastic “rudimentary” right ventricle
- Aorta and subaortic region may be hypoplastic
- Aortic arch may have hypoplasia, coarctation or complete interruption

e. Clinical presentation

i) Signs and symptoms

- Cyanosis at birth
- Tachypnea
- Poor feeding

f. Associated lesions

i) Patent foramen ovale (PFO), atrial septal defect (ASD)

ii) D-loop transposition of great arteries

iii) Left superior vena cava (LSVC) to coronary sinus

iv) Ventricular septal defect

v) Pulmonary valve stenosis

vi) Pulmonary atresia

vii) Coarctation of the aorta, associated with transposed great arteries

viii) Juxtaposition of the atrial appendages

g. Echocardiographic evaluation

i) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess flow across patent foramen ovale (PFO)/atrial septal defect (ASD), superior and inferior vena cava return to the right atrium, great artery relationship, ventricular septal defect (VSD) size and location

ii) Subcostal right anterior oblique view vertex down) to assess right ventricular outflow tract (RVOT), and pulmonary artery

iii) Apical 4-chamber view (vertex down) to assess left atrium and left ventricle, right ventricle (RV), ventricular septal defect (VSD), outflow tracts, and great arteries

iv) Parasternal long and short axis (vertex up) to assess ventricular septal defect(s), confirm relationship of great arteries, assess left ventricular systolic function, evaluate right ventricular outflow tract, pulmonary artery, and branch pulmonary arteries

v) Suprasternal short axis, high left parasternal view (vertex up) to assess branch pulmonary arteries, and patent ductus arteriosus

vi) Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

h. Treatment

i) Staged palliation

- First stage
  - Unrestricted pulmonary outflow- pulmonary artery (PA) banding
  - Severe pulmonary valve stenosis or pulmonary atresia: modified Blalock-Thomas-Taussig shunt (BTTS)
  - If coarctation of the aorta present, repair
  - If significant sub-aortic obstruction is present (transposition of the great arteries), may require Damus-Kaye-Stansel anastomosis
- Second stage
  - Bidirectional Glenn anastomosis
- Third stage
  - Modified Fontan procedure (fenestrated and non-fenestrated)

i. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate total cavopulmonary anastomosis for evidence of obstruction or thrombus

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

iii) Evaluate ventricular function

4. Hypoplastic left heart syndrome

a. Etiology

i) Describes a collection congenital heart anomalies in which the left ventricle is incapable of providing adequate systemic perfusion

ii) Include variations from mitral stenosis to atresia, left ventricular hypoplasia, aortic valve stenosis to atresia, and ascending aortic hypoplasia

iii) Often associated with coarctation of aorta

iv) 0.103 -0.267:1000 live births

v) May present with small patent foramen ovale/atrial septal defect or intact atrial septum (poor prognosis)

b. Pathophysiology

i) Postnatal survival depends upon patency of the ductus arteriosus, which supplies the systemic circulation

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

ii) Surgical palliation performed within the first week of life (stage 1 Norwood procedure)

iii) Unobstructed interatrial communication must be maintained

iv) 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 congestive heart failure (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)

d. Echocardiographic features (pediatric and adult patients)

i) Single, dominant RV

ii) Small LA

iii) Mitral valve hypoplasia or atresia

iv) Aortic valve hypoplasia or atresia

v) Varying degrees of left ventricular hypoplasia

vi) May present with no definitive LV cavity

vii) LV may have endomyocardial fibroelastosis (EFE)

viii) Varying degrees of ascending and aortic arch hypoplasia

ix) Coarctation of the aorta

e. Echocardiographic evaluation (newborn/unpalliated patients)

i) Subcostal coronal, left anterior oblique views (vertex down) to assess size and flow through patent foramen ovale (PFO)/atrial septal defect (ASD)

ii) Subcostal sagittal views (vertex down) to assess abdominal aortic flow, systemic venous return, ventricular function

iii) Apical 4-chamber view (vertex down) to assess right ventricular function, evaluate presence/severity of tricuspid valve regurgitation, demonstrate the degree of mitral stenosis/atresia, aortic valve stenosis/atresia, assess pulmonary venous flow

iv) Parasternal long (vertex up) to assess right atrium, right ventricle, and tricuspid valve, assess left side structures for size and patency

v) Parasternal short axis (vertex up) evaluate coronary artery origins, patent ductus arteriosus, right ventricular function

vi) Suprasternal short axis, high left parasternal view (vertex) to assess branch pulmonary arteries, and patent ductus arteriosus

vii) Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

f. Treatment

i) Staged surgical palliation

- "Hybrid" Norwood/modified Norwood palliation
- Glenn (superior cavopulmonary anastomosis)
- Fontan (inferior cavopulmonary baffle/conduit)
- Tricuspid repair/replacement
- Heart transplantation

g. 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 tricuspid regurgitation

v) Evaluate ventricular function

vi) Evaluate systemic inflow and outflow for obstruction

vii) Evaluate cavopulmonary anastomoses/conduits to ensure patency

J. Ventricular Anomalies

1. Ventricular septal defect (VSD)

a. Etiologies

i) Perimembranous

- Most common type of ventricular septal defect (VSD)
- Failure of membranous portion of ventricular septum to develop

- Located in membranous/perimembranous area of ventricular septum, in the left ventricular outflow tract below the aortic valve
- Smaller/restrictive defects associated with aortic valve prolapse/aortic valve regurgitation

ii) Inlet muscular

- Deficiency of inlet portion of ventricular septum
- Seen with atrioventricular valve septal defects
- Located in the inflow portion of the ventricular septum
- Isolated defects associated with straddling mitral or tricuspid valve

iii) Trabecular (muscular)

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

iv) Doubly committed juxta-arterial

- 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
  - Conal septum malaligned anteriorly creating right ventricular outflow tract obstruction (RVOTO)
  - Varying degrees of associated pulmonary valve, main pulmonary artery, and branch pulmonary artery hypoplasia
- Posterior malalignment
  - Posterior displacement of conal septum causing left ventricular outflow tract obstruction (LVOTO)
  - Associated with muscular aortic stenosis and/or aortic arch anomalies (interrupted aortic arch)

b. Pathophysiology

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

ii) Increased pulmonic flow

iii) Left atrial enlargement

iv) Left ventricular volume overload

- Left ventricular hypertrophy
- Elevated left ventricular end-diastolic pressure (LVEDP)
  - Elevated left atrial pressure
  - Elevated pulmonary venous pressure
- Enlarged left ventricle

c. Clinical presentation (pediatric and adult patients)

i) Signs and symptoms
- Small defects (restrictive)
- May have no hemodynamic significance
- High velocity jet with loud harsh murmur
- Subaortic defects associated with aortic valve prolapse and varying degrees of aortic regurgitation

ii) Large defects (unrestrictive)
- Cardiomegaly on x-ray
- Congestive heart failure
- Low frequency pansystolic murmur at lower left sternal edge or no systolic murmur

iii) 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 views

i) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess size and location of ventricular septal defect

ii) Subcostal right anterior oblique (vertex down) to assess ventricular septal defect and pulmonary artery
- Helpful view in tetralogy of Fallot to assess the ventricular septal defect and location and degree of pulmonary stenosis

iii) Apical 4-chamber view (vertex down) to assess cardiac size and function, evaluate LA and LV size and LV function, apical 5-chamber, evaluate for possible aortic valve prolapse and associated aortic valve insufficiency

iv) Parasternal long (vertex up) to assess ventricular septal defect

v) Parasternal short axis (vertex up) evaluate cardiac function, assess ventricular septal defect size and location
- Perimembranous ventricular septal defects seen at the level of the aortic valve in the 9 -11 o'clock position
- Muscular ventricular septal defects will be seen below the level of the aortic valve and in the muscular septum
    - Utilize slow sweeps with color

- Doubly committed juxta-arterial ventricular septal defects seen at the level of the aortic valve in the 1 o'clock position

f. Treatment

i) Small (restrictive) ventricular septal defects

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

ii) Large (unrestricted) ventricular septal defects

- Pericardial or synthetic patch closure
- Intraventricular baffle-type for double outlet right ventricle (DORV)
- If Eisenmenger syndrome present, ventricular septal defect (VSD) should not be closed

g. Echocardiographic assessment postoperatively

i) Evaluate for residual shunt

ii) Evaluate for additional (undetected) ventricular septal defects (VSD)

iii) Evaluate for aortic regurgitation and/or tricuspid valve regurgitation

iv) Evaluate for pulmonary hypertension

v) Assess outflow tract obstruction

2. Ventricular inversion (congenitally corrected transposition of the great arteries [CCTGA])

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 pulmonary artery

ii) Associated lesions

- Perimembranous ventricular septal defect
- Ebstein anomaly of the morphologic (systemic) ventricle
- Right ventricular outflow tract obstruction (RVOTO)
- Congenital complete heart block

b. Pathophysiology

i) Normal hemodynamic physiology

ii) Morphologic right ventricle - systemic cardiac output

iii) Morphologic left ventricle - pulmonary cardiac output

iv) Right ventricle remodels to accommodate systemic pressure

v) Left ventricle atrophies under pulmonary pressure

vi) Ebsteinoid tricuspid valve can lead to malcoaptation and regurgitation

vii) Severe volume load on right ventricle can cause congestive heart failure (CHF) in infancy

viii) Patients with complete heart block have higher mortality

ix) Right ventricular 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 congenital heart disease

ii) Congestive heart failure (CHF) caused by tricuspid regurgitation and/or systemic (right) ventricular failure

iii) Development of complete heart block

d. Echocardiographic features (pediatric and adult patients)

i) Apical 4 chamber view demonstrates attachment of left-sided atrioventricular valve (tricuspid) valve slightly apical to right-sided atrioventricular valve (mitral) valve

ii) Apical long 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 views

i) This lesion can be confusing; it is helpful to draw a picture of the patient's anatomy, to use as a guide while scanning

ii) Subcostal coronal and left anterior oblique views (vertex down) to assess for atrial level defect, left ventricular outflow tract obstruction (LVOTO), and ventricular septal defects

iii) Subcostal sagittal views (vertex down) to assess ventricular function

iv) Apical 4-chamber view (vertex down) to assess confirm ventricular arrangement, cardiac function, tricuspid valve morphology, function and competency, assess outflow tracts

v) Parasternal long (vertex up) to demonstrate features of left-sided morphologic right ventricle, image septal attachments of morphologic tricuspid valve, confirm ventriculo-infundibular fold separating tricuspid valve and aortic valve, assess outflow tracts for obstruction

vi) Parasternal short axis (vertex up) confirm ventricular morphology, evaluate ventricular sizes and systolic function, assess for ventricular septal defect(s)

f. Treatment

i) No treatment necessary for asymptomatic cases with no additional congenital heart disease (CHD)

ii) Moderate to large unrestrictive ventricular septal defect - primary closure

iii) Relief of pulmonary/subpulmonary stenosis if present

iv) Patients with ebsteinoid tricuspid valve and significant tricuspid regurgitation - pulmonary artery banding

v) Complete heart block - pacemaker insertion

vi) "Double switch" operation

- Senning or mustard (atrial switch) operation
- Jatene (arterial switch) operation

- Re-routes blood flow correctly; left ventricle receives pulmonary venous return and pumps to aorta, right ventricle receives systemic venous return and pumps to pulmonary artery

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) This lesion can be confusing; it is helpful to draw a picture of the patient's anatomy, post-surgery to use as a guide while scanning

ii) Evaluate systemic ventricular (right ventricle) function (all patients)

iii) Assess presence and degree of tricuspid regurgitation

iv) Evaluate for residual ventricular septal defect- status post repair

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

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

K. Semilunar Valve Anomalies

1. Aortic stenosis (AS)

a. Types and etiology

i) Subvalvular tunnel

- Hypoplastic left ventricular outflow tract

ii) Left ventricular outflow tract obstruction (LVOTO) associated with hypertrophic cardiomyopathy

- Systolic anterior motion of mitral valve chordae and apparatus causing left ventricular outflow tract obstruction (LVOTO)
- May be isolated mid-cavitary

iii) Discrete subaortic membrane

- Associated with
  - Ventricular septal defect (VSD) or spontaneously closed VSD
  - Shone's complex
- Fibrous or fibromuscular tissue in left ventricular outflow tract (LVOT)
  - Circumferential or partial within LVOT

iv) Bicuspid aortic valve

- Incidence 2% of population
- Associated with coarctation of the aorta
- Congenitally malformed leaflets

v) Congenital aortic valve stenosis

- Three leaflets
- Varying degrees of commissural fusion

vi) Unicuspid aortic valve

- Congenitally malformed leaflets
- Single slit-like commissure in cross-section

vii) Quadricuspid aortic valve

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

viii) Supravalvular aortic stenosis

- 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 from left ventricle (LV)

ii) Concentric left ventricular hypertrophy (LVH) develops

- Calculation of LV mass assists with evaluating the left ventricular workload

iii) Left atrial enlargement may also occur in patients with LVH

- Associated with abnormal left ventricular diastolic function

c. Clinical presentation

i) Signs and symptoms

- More common in males than females
- Dependent upon severity (may be asymptomatic)
- Critical AS
  - Low cardiac output
  - Reduced ejection fraction
  - Loud systolic murmur
  - Congestive heart failure
  - Shock
  - Dependent on a patent ductus arteriosus (PDA)

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 and root, sinotubular junction, and ascending aorta

e. Echocardiographic views

i) Subcostal left anterior oblique views (vertex down) to assess left ventricular outflow tract, aortic valve, and proximal ascending aorta

ii) Apical 4-chamber view (vertex down) to assess cardiac function, presence of left ventricular hypertrophy

iii) Apical 5-chamber view (vertex down) to assess left ventricular outflow tract, aortic valve, and proximal ascending aorta

iv) Parasternal long (vertex up) to assess left ventricular outflow tract, and aortic valve cusp appearance and mobility

v) Parasternal short axis (vertex up) evaluate aortic valve cusps and valve function, assess for left ventricular hypertrophy

vi) High right parasternal sagittal view to assess ascending aorta

- View provides an optimal spectral Doppler angle

vii) Pedoff transducer (adolescents to adults)

- Right parasternal window
- Suprasternal notch window
- Apical 5-chamber window

f. Treatment

i) Catheterization

- Balloon dilation

ii) Surgery

- Valve repair
- Ross procedure
- Valve replacement

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate transvalvular pressure gradient

ii) Evaluate for aortic valve regurgitation

iii) Evaluate for prosthesis stenosis/failure/paravalvular leak

iv) Evaluate left ventricular function

v) Assess ascending aorta and arch

vi) Evaluate right ventricle to pulmonary artery connection and branch pulmonary arteries (Ross procedure)

2. Pulmonary stenosis

a. Types and etiology

i) Pulmonary valve stenosis

- Bicommissural pulmonary valve
- Tri-leaflet valve with cusp fusion

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 pulmonary artery (PA) segment
- Branch pulmonary artery (PA) stenosis

iv) Subvalvular pulmonary stenosis

- Muscle bundle at the infundibular opening, creating subvalvular obstruction

b. Pathophysiology

i) Increased resistance to flow from right ventricle (RV)

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 pulmonary valve stenosis

- Cyanosis

d. Echocardiographic features

i) Thickened leaflets

ii) Doming of leaflets

iii) RVH

iv) Doppler pressure gradient

v) Evaluate for infundibular stenosis

e. Echocardiographic views

i) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess right ventricle size and function, right ventricular outflow tract, and pulmonary valve size and leaflet motion

ii) Subcostal right anterior oblique (vertex down) to assess right ventricular outflow tract, pulmonary valve, and main pulmonary artery

- Helpful view in assessing the location and degree of pulmonary stenosis

iii) Apical 4-chamber view with anterior sweep (vertex down) to assess cardiac function, right ventricular outflow tract and pulmonary valve

iv) Parasternal long (vertex up) to assess right atrium, right ventricle, tricuspid valve, and pulmonic valve

v) Parasternal short axis (vertex up) evaluate right ventricular outflow tract, pulmonic valve, main and branch pulmonary arteries

f. Treatment (pediatric and adult patients)

i) Catheterization

- Balloon dilation

ii) Surgery

- Pulmonary valve repair

- Transannular patch (annular hypoplasia)
- Non-transannular patch (right ventricular outflow tract obstruction, infundibular hypoplasia)
- Pulmonary valve replacement (adolescents and adults)

g. Post-operative/repair evaluation

i) Evaluate trans-valvular pressure gradient

ii) Evaluate for residual pulmonary valve stenosis, presence/severity of pulmonary valve regurgitation

iii) Assess for residual right ventricular outflow tract obstruction

iv) Evaluate right ventricular size and function

v) Assess tricuspid valve regurgitation

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 right ventricular outflow tract and pulmonary artery without a ventricular septal defect

- Two morphologic types
  - Hypoplastic right ventricle and tricuspid valve, with right ventricular hypertrophy, and coronary artery fistulae/sinusoids
  - Dilated thin-walled right ventricle, severe tricuspid valve regurgitation, severely dilated right atrium

b. Pathophysiology

i) Systemic venous return crosses PFO (right to left flow); mixing in left atrium causes cyanosis in newborn

ii) Pulmonary blood flow is dependent on ductus arteriosus (ductal-dependent lesion)

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

iv) Pulmonary artery flow is ductal dependent at birth unless major aorto-pulmonary collateral arteries (MAPCAs) are present (disease variant)

c. Clinical presentation

i) Cyanosis at birth

ii) May present with congestive heart failure (CHF), hepatomegaly in subtype with massive tricuspid regurgitation and right heart enlargement

iii) Adolescents and adults may present with congestive heart failure (CHF) due to poor LV function and development of mitral valve regurgitation

iv) Myocardial infarction (MI) may occur due to presence of right ventricular 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 views

i) Subcostal coronal, left anterior oblique, and sagittal views to assess systemic venous return, atrial septal defect, right atrium/right ventricle size and function

ii) Subcostal right anterior oblique (vertex down) to assess inflow and outflow of the right heart

- Helpful view to assess right ventricular outflow tract for antegrade flow, pulmonary artery size, and presence of a patent ductus arteriosus

iii) Apical 4-chamber view (vertex down) to assess cardiac function, assess right ventricle for sinusoids, assess tricuspid valve

iv) Parasternal long (vertex up) to assess right atrium, right ventricle, and tricuspid valve

v) Parasternal short axis (vertex up) evaluate coronary artery anatomy and flow, assess right ventricular outflow tract for antegrade flow, presence of patent ductus arteriosus, assess branch pulmonary arteries for continuity

vi) Suprasternal short axis, high left parasternal view (vertex up) to assess branch pulmonary arteries, and patent ductus arteriosus

f. Treatment

i) Staged approach needed in most cases

- Ductal stenting (stage 1)
- Systemic-to-pulmonary (Blalock-Thomas-Taussig shunt (BTTS)) with patent ductus arteriosus ligation (stage 1)
- Glenn (stage 2)
- Fontan (stage 3)

ii) Pulmonary valve perforation and balloon dilation possible in cases with adequate tricuspid valve and right ventricular size and function

iii) Heart transplantation may be necessary in cases with right ventricular dependent coronary circulation (RVDCC)

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Assess left ventricular function

ii) Evaluate atrial shunting

iii) Assess presence and severity of mitral regurgitation

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)

- Right ventricular outflow tract and degree of regurgitation (if valvotomy performed)

L. Aortic Arch Anomalies

1. Coarctation of the aorta

a. Types

i) Pre-ductal (arch hypoplasia)

ii) Juxta-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 through the left heart during embryologic and fetal development

iii) Environmental factors

iv) Occurs in 5-7% of all congenital heart disease (CHD)

v) Associated lesions

- Shone's complex
  - Bicuspid aortic valve
  - Left superior vena cava to the coronary sinus
  - Mitral valve abnormalities
- D-transposition of the great arteries (DTGA)

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 left ventricle
  - Increased left ventricular end diastolic pressure
  - Pulmonary edema

ii) Signs and symptoms in adolescence and adulthood

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

e. Echocardiographic features

i) 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)

ii) Post-stenotic dilation may be present

iii) Increased velocity seen on spectral Doppler in area of coarctation

iv) Doppler waveforms in abdominal aorta may be blunted with low velocity and continuous antegrade diastolic flow

v) Color Doppler will reveal turbulent flow in stenotic region

vi) LVH

vii) LV function may be decreased

viii) Signs of elevated LVEDP

- Left atrial enlargement
- Monophasic mitral valve inflow pattern

f. Echocardiographic views

i) Subcostal sagittal view (vertex down or up) focused clip of abdominal aorta, spectral Doppler performed at the level of the diaphragm

ii) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess atrial level shunt, cardiac function

iii) Apical 4-chamber view (vertex down) to assess cardiac function

iv) Apical 5-chamber view (vertex down) to assess left ventricular outflow tract and flow velocities across aortic valve

v) Parasternal long (vertex up) to assess and measure the aortic valve annulus, sinuses, sinotubular junction, and ascending aorta

vi) Parasternal short axis (vertex up) evaluate aortic valve morphology, cardiac function, presence of patent ductus arteriosus

vii) Suprasternal short axis, to evaluate aortic arch sidedness, assess presence of patent ductus arteriosus, and evaluate the descending aorta

viii) Suprasternal long axis, high left parasternal views (vertex up) to assess aortic arch and patent ductus arteriosus

- Slow sweeps and view modification are required to pinpoint the location of the coarctation

- Use higher frequencies for better temporal resolution

g. Treatment

i) Surgery

- End-to-end anastomosis (discrete narrowing)
- Extended end-to-end anastomosis (diffuse hypoplasia)

ii) Catheterization

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

h. 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 atrioventricular or mitral valve 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 ventricular septal defect
- Subaortic stenosis
- Left superior vena cava to a dilated coronary sinus
- Mitral valve abnormalities
- Hypoplastic left heart syndrome

c. Pathophysiology

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

ii) Ascending aorta provides coronary and cerebral perfusion

iii) Posterior malalignment ventricular septal defect is most commonly 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) Repaired patients may present with symptoms of re-coarctation

e. Echocardiographic views

i) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess left ventricular outflow tract and proximal ascending aorta

ii) Apical 5-chamber view (vertex down) to assess left ventricular outflow tract, ventricular septal defect, and presence and degree of subaortic narrowing

iii) Parasternal long (vertex up) to assess left ventricular outflow tract, measure the aortic valve annulus, sinuses, sinotubular junction, and ascending aorta, assess ventricular septal defect and presence and degree of subaortic narrowing

iv) Parasternal short axis (vertex up) to evaluate aortic valve morphology, patent ductus arteriosus

v) Suprasternal short axis, (vertex up) to evaluate aortic arch sidedness, assess presence of patent ductus arteriosus, and evaluate brachiocephalic vessels to determine type of interruption (depending on branching pattern)

vi) Suprasternal long axis (vertex up) to evaluate brachiocephalic vessels to determine type of interruption, assess aortic arch, and patent ductus arteriosus

f. Treatment

i) Aortic arch repair

- Excision of ductal tissue with end-to-side arch anastomosis

ii) Ventricular septal defect closure

iii) Resection of obstructive subaortic 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 ventricular septal defect baffle with right ventricle to pulmonary artery homograft

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate arch for residual obstruction

ii) Assess left ventricular outflow tract for obstruction

iii) Assess for residual ventricular septal defect(s)

iv) Evaluate ventricular function

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

M. 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, infant of diabetic mother (IDM)

iii) Environmental factors may play a role

iv) 10th most common congenital heart disease (CHD)

v) 2nd most common cyanotic lesion

c. Pathophysiology of D-TGA

i) Two parallel circuits exist

ii) Systemic venous blood returns to the right ventricle and then is recirculated to the systemic circulation through the aorta

iii) Pulmonic venous blood returns to the left ventricle and then is recirculated to the pulmonary circulation through the pulmonary artery

d. Clinical presentation D-TGA

i) Cyanotic at birth

ii) Single loud second heart sound

iii) Ventricular septal defects/left ventricular outflow tract obstruction variants will have flow murmur

iv) Coarctation variants may have significant metabolic acidosis

e. Associated lesions

i) Ventricular septal defects in 40-45 % of patients

ii) Left ventricular outflow tract obstruction in 20-30%

iii) Coarctation of the aorta

iv) Systemic venous anomalies

f. Echocardiographic features

i) Ventriculo-arterial discordance

ii) Fibrous continuity between the mitral and PV

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 views

i) Subcostal coronal and left anterior oblique views (vertex down) to assess atrial level communication, slow sweep will demonstrate the first great vessel arising the from the left ventricle and bifurcating, continuous with a pulmonary artery, assess for ventricular septal defect(s), and outflow tract obstruction

ii) Subcostal sagittal views (vertex down) will demonstrate a parallel relationship of the great arteries

iii) Apical 4-chamber view (vertex down) with posterior to anterior sweep will confirm the ventriculo-arterial discordance, assess cardiac function, assess for ventricular septal defect(s), and outflow tract obstruction

iv) Parasternal long axis (vertex up) will demonstrate the parallel relationship of the great arteries, assess left ventricular outflow tract, and ventricular septal defects

v) Parasternal short axis (vertex up) evaluate semilunar valve morphology, coronary artery origins and proximal courses, patent ductus arteriosus

vi) Suprasternal short axis, high left parasternal view (vertex up) to assess branch pulmonary arteries, and patent ductus arteriosus

vii) Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

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 (dextro transposition of the great arteries (D-TGA); double outlet right ventricle (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
- Evaluate coronary artery anastomoses using color Doppler
- Evaluate semilunar valves for stenosis and/or regurgitation
- Assess ventricular function

ii) Rastelli

- Evaluate baffle for residual shunts
- Evaluate left ventricle to aorta pathway
- Assess right ventricle to pulmonary artery connection
- Assess atrioventricular valve competency
- Evaluate ventricular function

iii) Mustard or Senning

- Evaluate atrial baffles for leaks or obstruction
- Assess ventricular function
- Assess tricuspid valve function and presence of regurgitation

2. Tetralogy of fallot (TOF)

a. Types

i) TOF with pulmonary stenosis (varying degrees)

ii) TOF with pulmonary atresia

iii) TOF with absent PV syndrome

iv) TOF with complete atrioventricular septal defect (AVSD)

b. Etiology

i) Occurs in 4-8% of all congenital heart disease (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
- Branch 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 ventricular septal defect result in ventricular-level right-to-left shunt and cyanosis

ii) Degree of cyanosis directly related to severity of pulmonary outflow obstruction

iii) Ductal patency improves pulmonary blood flow but may contribute to pulmonary overcirculation 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 tetralogy of fallot (TOF) with Absent pulmonary valve syndrome usually have massively dilated PA branches causing airway compression, exacerbating cyanosis

ii) May present with over-circulation and signs of congestive heart failure (“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) Short axis view cases of TOF with absent PV may have right heart enlargement due to significant associated pulmonary regurgitation

ii) Abnormalities of aortic arch sidedness and branching may be present

f. Echocardiographic views

i) Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess size and location of ventricular septal defect

ii) Subcostal right anterior oblique (vertex down) to assess ventricular septal defect and pulmonary artery

- Helpful view in tetralogy of Fallot to assess the ventricular septal defect, location and degree of pulmonary stenosis
- Right heart inflow-outflow view

iii) Apical 4-chamber view (vertex down) with anterior sweep to assess the ventricular septal defect, right ventricular outflow tract, right ventricular outflow tract and pulmonary artery

iv) Parasternal long (vertex up) to assess classic features of TOF, aortic override of large anterior malalignment ventricular septal defect, and right ventricular hypertrophy (RVH)

v) Parasternal short axis, high parasternal views (vertex up) demonstrate anteriorly deviated conal septum with infundibular narrowing, and varying degrees of pulmonic valve annular hypoplasia, assess branch pulmonary artery relationship and size, assess for coronary artery anomalies

- TOF associated with conal branches near or crossing the right ventricular outflow tract from the right coronary artery

vi) Suprasternal short axis, evaluate aortic arch sidedness, and branching pattern

vii) Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

g. Treatment

i) Repair dependent on anatomy

- May need staged approach if anatomy unfavorable for repair
  - Blalock-Taussig-Thomas shunt (BTTS) early
  - Complete repair involving ventricular septal defect (VSD) closure and relief of pulmonary stenosis (around) 6-months of age

ii) Transannular patch repair

iii) Non-transannular repair

iv) Complete repair with right ventricle to pulmonary artery (RV-PA) homograft/conduit

v) Complete repair with plication of main pulmonary artery and branch pulmonary arteries (TOF, absent pulmonary valve)

vi) Adolescent and adult patients may need revision of RV-PA connection

- Stent placement(s) for residual obstructive right ventricular outflow tract and pulmonary artery lesions
- Re-operative surgical pulmonary valve replacement
- Percutaneous valve placements

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate aorta-to-pulmonary shunt

ii) Evaluate for residual ventricular septal defects

iii) Assess right ventricular outflow tract for residual obstruction and possible aneurysm formation

iv) Evaluate pulmonic valve for residual stenosis or regurgitation

v) Evaluate pulmonary arteries for residual narrowing or dilation

vi) Evaluate tricuspid valve regurgitation

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 ventricular septal defect

- Relationships of the great arteries
  - Aorta is to the right and posterior to the pulmonary artery
  - Aorta is right and lateral to the pulmonary artery
  - Aorta is right and anterior to the pulmonary artery
  - Aorta is left and anterior to the pulmonary artery
- 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 congenital heart disease (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 septal defect (AVSD)
- Aortic arch anomalies
  - Coarctation
- Arch branching malformations
- Systemic venous anomalies

c. Pathophysiology

i) Highly variable depending on ventricular septal defect size, location, outflow tract placement and patency and the streaming characteristics of blood flow

- Ventricular septal defect physiology (pulmonary overcirculation)
- Tetralogy of Fallot physiology (varying degrees of cyanosis)
- D-transposition of the great arteries 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
- Congestive heart failure (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 atrioventricular valves and semilunar valves (bilateral conus)

f. Echocardiographic views

i) Subcostal coronal and left anterior oblique views (vertex down) to assess atrial level communication, slow anterior sweep will demonstrate the great artery relationship, assess ventricular septal defect(s), and outflow tract obstruction

ii) Subcostal sagittal views (vertex down) will demonstrate a parallel relationship of the great arteries

iii) Apical 4-chamber view (vertex down) with posterior to anterior sweep will demonstrate the relationship of great arteries, and ventricular septal defect, assess for outflow tract obstruction

iv) Parasternal long axis (vertex up) will demonstrate the parallel relationship of the great arteries, assess outflow tracts, and ventricular septal defect(s)

v) Parasternal short axis (vertex up) evaluate great artery relationship

vi) Suprasternal short axis, high left parasternal view (vertex up) to assess aortic arch sidedness, branch pulmonary arteries, and patent ductus arteriosus

vii) Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

g. Treatment

i) Complete repair

- Baffle closure of ventricular septal defect connecting the left ventricle to the nearest semilunar valve
- Rastelli procedure
- Nikaidoh procedure
- Arterial switch operation with ventricular septal defect closure

ii) Staged single ventricle palliation for cases with

- Straddling atrioventricular valves
- Remote ventricular septal defect
- Hypoplastic ventricles

h. Echocardiographic assessment postoperative

i) Evaluate great artery anastomosis sites for obstruction

ii) Evaluate for residual ventricular septal defect

iii) Assess outflow tracts for residual obstruction

iv) Evaluate pulmonary arteries

v) Evaluate for tricuspid valve regurgitation

vi) Assess ventricular function

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. Myocardial oxygen supply/demand mismatch
    - b. Perfusion abnormalities
    - c. Diastolic dysfunction
    - d. Systolic dysfunction
    - e. Increased LV filling pressures
    - f. Ischemic ECG changes
    - g. Symptoms (angina, dyspnea)
  - 3. Indications/clinical applications
    - a. CAD diagnosis and prognosis
    - b. Myocardial viability
    - c. Pulmonary hypertension
    - d. Valvular heart disease
    - e. HCM
    - f. Diastolic dysfunction
    - g. Pre-operative risk stratification
  - 4. Contraindications
    - a. Acute coronary syndromes
    - b. Severe cardiac arrhythmias
    - c. Severe hypertension
    - d. Symptomatic severe aortic stenosis
  - 5. Safety
  - 6. Diagnostic accuracy
- B. Types of Stress Echocardiography
  - 1. Exercise stress echocardiography

a. Patient preparation and ECG-lead placement
b. Indication
c. Relative contraindications
d. Stress testing system
    i) Treadmill
    ii) Bicycle
    iii) Other
e. 12-lead ECG system
f. Blood pressure device
g. Emergency equipment
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
4. Hemodynamic

C. Corresponding Perfusion Zones of the Major Coronary Arteries
    1. LAD

2. LCX
3. RCA

D. 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. Imaging during the study (pharmacologic stress or bicycle stress)
4. Post-exercise echocardiographic images (treadmill)
   a. Time limitations
   b. Patient position
   c. Time increments
5. Digital image acquisition
6. UEA for LV opacification or perfusion

E. 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

F. 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

G. 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. Patient preparation
    e. Patient sedation
    f. Laboratory set up /clean up
    g. 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: 3D Echocardiography

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

XVII. REAL TIME 3-D ECHOCARDIOGRAPHY

- A. Imaging Methods and Display
  - 1. Image acquisition modes
    - a. Multiplane imaging
    - b. Real-time or live 3D imaging
    - c. ECG-gated multi-beat acquisition (full volume or zoom mode)
  - 2. Color Doppler data sets
    - a. Superimpose flow velocity data onto a 3D zoom or full-volume data set
    - b. Challenging – narrower sector widths and lower frame rates
- B. Clinical Indications
  - 1. Left and right ventricular assessment
    - a. Function
    - b. Volume/mass
    - c. Eliminates geometric assumptions and diminishes foreshortening
  - 2. Valvular evaluation
    - a. Quantitative assessment of mitral stenosis
    - b. Visualization of the tricuspid valve
    - c. TEE-based imaging of the aortic valve
  - 3. Left atrial appendage
  - 4. Use in interventional imaging
- C. Modality Comparison
  - 1. Validation against cardiac MRI
  - 2. Advantages compared to 2D echocardiography
  - 3. Limitations/pitfalls

Section XVIII: Enhanced Cardiac Ultrasound

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

XVIII. ENHANCED CARDIAC ULTRASOUND

- A. Agitated saline
  - 1. Indications and contraindications
  - 2. Clinical applications
  - 3. Set up and supplies
  - 4. IV insertion techniques
- B. Transpulmonary Agents
  - 1. Indications and contraindication
  - 2. Clinical applications
- C. Set Up and Supplies
  - 1. Bubble characteristics
    - a. Size
    - b. Stability
    - c. Composition
    - d. Acoustic properties
      - i) Acoustic impedance
      - ii) Resonant frequency
  - 2. Technical criteria
    - a. Appropriate rate and concentration
      - i) Bolus
      - ii) Diluted bolus
      - iii) Continuous infusion
      - iv) Set up and supplies
        - Resting TTE
        - Stress
        - TEE
    - b. Image acquisition
      - i) Continuous
      - ii) Sequential
      - iii) Gated
    - c. Appropriate equipment settings/optimization
      - i) Mechanical index (MI)

- High, intermediate, low

ii) Transducer frequency

iii) Harmonics

- Dynamic range
- Receiver filter
- Transmit frequency spectrum

iv) Pulse inversion

v) Power modulation

vi) Power pulse inversion

vii) Focal zone location

viii) Compression/dynamic range

ix) Gain

x) Effects of inappropriate settings

- Artifacts
- Attenuation
- Blooming
- Swirling

xi) Indications

- Suboptimal examination criteria

xii) Contradictions

d. Applications

i) Endocardial border definition

ii) Myocardial perfusion imaging

- Low MI (LMI)
- Very low MI (VLMI)

iii) Doppler signal enhancement

iv) Definition of cardiac mass

v) Left ventricular ejection fraction/function

vi) Left ventricular non-compaction

vii) Apical hypertrophic cardiomyopathy

e. Informed consent

f. Patient monitoring/safety

i) Allergic/anaphylactoid reactions

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

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
  - 3. Structural heart interventions
- E. Speckle Tracking Echocardiography (STE)
  - 1. Definition
    - a. Positive strain
    - b. Negative strain
    - c. Color strain tracking
    - d. Units
  - 2. Clinical application
  - 3. Technical consideration
- F. Short-Term Mechanical Circulatory Support
  - 1. Devices
    - a. Intra-aortic balloon pump
    - b. Extra-corporeal membrane oxygenation (ECMO)
    - c. TandemHeart
    - d. Impella
  - 2. Imaging considerations
- G. Left Ventricular Assist Devices
  - 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 (closed loop 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) 2D/3D/M-mode 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

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) Pseudo infarction pattern
   b. Bone marrow biopsy
6. Common treatment
   a. Medical
      i) Supportive treatment of cardiac failure
      ii) Chemotherapy
   b. Surgical
      i) Liver transplant (TTR amyloid)
      ii) Stem cell transplant
      iii) 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

g. Strain pattern of apical sparing

5. Related testing

a. Labs: persistent (at least six months) increase in eosinophil count > 1500 cells/mm^3 or documented on at least two occasions

6. Common treatment

i) Medical

ii) Treat CHF

iii) Anticoagulants

iv) Treat underlying hypereosinophilia (steroids)

7. 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. 70% of cases reported in patients aged 25-45
4. Echocardiographic features
   a. Most important manifestation is caused by pulmonary involvement
      i) Right-sided heart failure
      ii) Pulmonary hypertension
   b. Enlarged LV with decreased systolic function
   c. Regional wall motion abnormalities
   d. Posterolateral wall thinning with or without aneurysm
   e. Diastolic dysfunction (precedes systolic)
   f. Valvular dysfunction
   g. Pericardial effusion
   h. Patchy strain pattern
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
      ii) Automatic implantable cardioverter-defibrillator
      iii) Ventricular aneurysm resection
      iv) 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
   f. May see reversal of LV dysfunction with successful treatment
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
        ii) Pericardial (most 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 (most common)

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. Granulomatosis with Polyangiitis

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

XXI. CARDIAC TRANSPLANTATION

- A. Common 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. 2D/3D
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
3. Strain
a. Reduced global LV and RV strain compared to normals

F. Echo Features of Rejection
1. Rejection associated with non-compliance of left ventricle due to extracellular infiltration and edema
2. Routine echo evaluations not recommended but may be helpful
3. Possible indicators of rejection
a. Increase in LV wall thickness, LV mass
b. Decrease in LV and/or RV systolic function
c. Increase in RVSP
d. New pericardial effusion
e. Decrease in global longitudinal strain
f. Restrictive filling

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

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
- 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. Transcatheter mitral valve replacement/repair - TEER
  - 4. Tricuspid valve intervention
  - 5. Perivalvular leak closure
  - 6. Transcatheter based shunt closure
    - a. ASD
    - b. VSD
  - 7. 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
CMR	Cardiac Magnetic Resonance
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
PAH	Pulmonary Arterial Hypertension
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
PH	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
RWMA	Regional Wall Motion Abnormality
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
SCAD	Spontaneous Coronary Artery Dissection
SPWMD	Septal to Posterior Wall Mechanical Delay
SSN	Suprasternal Notch
STE	Speckle Tracking Echocardiography
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

U

UEA	Ultrasound Enhancing Agent

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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