National Education Curriculum

Specialty Curricula

Congenital 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. Compare and contrast the anatomy of aorta and vena cava
4. Describe the anatomy of the pericardium, the myocardium, and endocardium
5. Explain the relational anatomy of the heart in the thoracic cavity
6. Describe the location of the apex in relationship to other structures in the heart
7. Explain how the body habitus influences the position of the heart in the thoracic cavity

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 (AVV)
      - i) Mitral valve (MV)

- Anterior leaflet
  - Scallops (A1, A2, A3)
- Posterior leaflet
  - Scallops (P1, P2, P3)
- Papillary muscles
  - Septophobic papillary muscles

ii) Chordae tendineae

iii) Annulus

iv) Tricuspid valve (TV)

- Septal leaflet
- Posterior leaflet
- Anterior leaflet

v) Papillary muscles

- Septophillic papillary muscles

vi) Chordae tendineae

vii) 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

- Sinotubular junction

iii) Ascending aorta

iv) Aortic arch

v) Descending aorta

vi) Abdominal aorta

d. Major branches

i) Brachiocephalic (innominate)

- Bifurcates 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)

3. Coronary arteries

a. Left coronary artery

i) Left anterior descending (LAD)

ii) Left circumflex (LCX)

b. Right coronary artery (RCA)

c. Congenital variations

d. Flow patterns

e. Flow reserve

4. Superior vena cava (SVC) and inferior vena cava (IVC)

5. Pulmonary veins

6. Coronary venous system

a. Coronary sinus

b. Differentiation from descending Ao

c. Atrioventricular groove

7. 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: 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

II: PRINCIPLES OF CARDIAC HEMODYNAMICS AND CARDIAC CYCLE

- 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 (Poiseuille's) 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
    - d. Poiseuille's law
      - i) Bernoulli principle
        - Conservation of energy
        - Bernoulli equation
          - Assumption
            - ~ Flow acceleration + viscous friction are negligible
            - ~ No energy transfer
      - ii) Modified Bernoulli equation
      - iii) Simplified Bernoulli equation
      - iv) Clinical use
        - Valvular stenosis (aortic, pulmonic)
        - PA pressure
      - v) Reynolds number

- Point at which turbulence occurs

C. Physiology and Hemodynamic Information Derived from Doppler

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

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

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

E. Cardiac Cycle
1. Electrocardiogram
    a. P wave
    b. QRS complex
    c. ST segment
    d. 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
- MS
- Increased ventricular end-diastolic pressure

c. Diastasis

d. Atrial systole (atrial contraction)

i) Atrial pressure exceeds ventricular pressure

ii) MV opens and late LV filling occurs

4. Ventricular systole

a. Isovolumic relaxation time

b. Isovolumic contraction time (IVCT)

c. Rapid ejection

d. Reduced ejection

e. Factors that influence ventricular systole

i) Ventricular function

ii) Afterload

iii) Preload

iv) Frank-Starling's law

F. Pulmonary Veins

1. Vein characteristics
2. Anatomic location
3. Doppler interrogation
4. Waveform characteristics

a. Systole

b. Diastole

G. Cardiac Catheterization

1. Clinical utility

a. Intracardiac pressure

b. Oxygen saturation

c. Coronary artery assessment

d. Other

2. Normal pressure tracings

a. Right heart

b. Left heart

Section III: Normal and Altered Electrical Activation

1. List the events that lead to myocardial contraction
2. Describe normal electrical pathways
3. Describe the location and function of each portion of the cardiac conduction system
4. Define normal electrical wave forms seen on ECG
5. Describe echocardiographic findings associated with altered electrical activation
6. Identify abnormal ECG tracings
7. Recognize abnormal ECG tracings

III: NORMAL AND ALTERED ELECTRICAL ACTIVATION

A. Normal Electrical Activation

1. Electrical pathways
   - a. Sinoatrial (SA) node
   - b. Intra-atrial tracts
   - c. Atrioventricular (AV) node
   - d. Bundle of His
   - e. Left and right bundle branches
   - f. 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 (PVC)
     - 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

- Mobitz I,
- Mobitz II

iii) Complete heart block

g. Pacemaker

i) Single chamber

ii) Dual chamber

iii) Bi-ventricular pacemakers

Section IV: Patient Preparation and Indications

1. Identify appropriate indications for ordering an echocardiogram in pediatric congenital cardiac patients
2. Review and interpret relevant patient history and current vital signs prior to the procedure
3. Describe how to prepare the exam room and select appropriate transducer options for pediatric patients
4. Demonstrate correct patient positioning techniques to optimize echocardiogram results
5. Apply Z score methods to echocardiographic measurements for pediatric assessment
6. Recognize the need for and administer sedation safely when indicated
7. Generate comprehensive exam reports that effectively communicate findings
8. List and describe the top warning signs for critical congenital heart defects in pediatric patients

IV: PATIENT PREPARATION AND INDICATIONS

- A. Review Patient History and Prior Exams
  - 1. Referral
  - 2. Prior cardiology visit note
  - 3. Prior echocardiograms and imaging reports
- B. Current Vital Signs
  - 1. Blood pressure
    - a. Blood pressure monitoring is measured on both right and left arms
    - b. If a difference of 12 mmHg between left and right arms is documented, then both right and left calf or thigh blood pressures should be taken and documented as well
  - 2. Pulse oxygen
    - a. Oxygen saturation (SpO^2) is measured in the right hand (preductal) and on either foot (post-ductal)
    - b. Normal is above 95%
    - c. Normal saturation for baffle is 93% and above
    - d. Post-ductal measurement of SpO^2 is important because it detects right-to-left shunting of desaturated blood through the ductus arteriosus
  - 3. Arrythmias/pacing noted on rhythm strip
- C. Symptoms
- D. Maternal history, if needed and available
  - 1. Past drug use
  - 2. Diabetes
  - 3. Cardiotoxic drug exposure
- E. Create an Inviting and Relaxing Environment in the Exam Room
  - 1. Include family in exam room
  - 2. Comfortable room temperature for patient
  - 3. Television with a variety of age-appropriate viewing options
  - 4. Toys
  - 5. Child life specialists
  - 6. Available pacifiers and blankets
  - 7. Utilize swaddling with the chest exposed in infants

F. Multiple Transducer Options to Provide Appropriate Penetration and Resolution for a Variety of Body Sizes

1. Neonatal
2. Infants
3. Small children
4. Adolescents
5. Adults
6. High BMI adults

G. Patient Positioning

1. Older patients will be left lateral decubitus
2. Neonates and infants can be scanned supine or rolled to the left side
3. Dextrocardia patients will be right lateral decubitus

H. Z Score Method Evaluation

1. Dubois and Dubois method
    a. BSA=0.00718 x height $^{0.725}$ x weight $^{0.425}$
2. The method of Dreyer and Ray
    a. BSA= 0.1 x weight 0.6666
3. Boyd published method
    a. BSA= 0.0004688 x (1000 x weight)0.8168-0.0154kg(1000 x weight)
    b. BSA= 0.0003207 x (1000 x weight)0.7285-0.0188kg(1000 x weight) x height 0.3

I. Sedation

1. May be required in situations when the patient cannot cooperate for the exam
2. Utilize facility protocols related to sedation administration and monitoring

J. Post Examination Reporting

1. Reports should include patient demographics, biometrics, vital signs, body surface area
2. Section detailing quantitative measurements of images and Doppler
3. Summary of normal findings
4. Conclusion of abnormal finding or changes
5. Recommendation made to refer to the IAC pediatric and adult congenital reporting template

K. Top Warning Signs of Critical Congenital Heart Disease

1. Cannot see the aortic arch
2. Retrograde flow in ascending aorta
3. Exclusive right to left shunting at the atrial or ductal level
4. Apical 4 chamber doesn’t look like an apical 4 chamber
5. An atrioventricular valve with moderate to severe regurgitation
6. Apex is pointed midline or rightward
7. Cannot demonstrate a normal parasternal long axis (PLAX) of the LV and Aorta in the same picture
8. There is no normal parasternal short axis (PSAX) or the RV cannot open into a PV/PA

9. Aortic valve and PV are parallel, cannot see them crisscross

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Section V: Radiation Safety

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

V: RADIATION SAFETY

A. Basic radiation principles

B. Basic radiation safety principles

C. Personal protection techniques

D. Regulatory issues

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Section VI: 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 formation
3. Describe the formation of the atrioventricular valves and the semilunar valves
4. Compare fetal and neonatal circulation

VI: BASIC EMBRYOLOGY

- 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
- B. Ventricular Looping
  - 1. Dextro (rightward) ventricular looping (D-loop)
  - 2. Levo (leftward) ventricular looping (L-loop)
- C. Regions of Heart Tube
  - 1. Sinus venosus
  - 2. Primitive atria
  - 3. Atrioventricular canal
  - 4. Primitive LV
  - 5. Interventricular foramen
  - 6. Primitive RV
  - 7. Conus arteriosus
  - 8. Truncus arteriosus
  - 9. Aortic sac
  - 10. Aortic arches
  - 11. Pulmonary arteries
- D. Septation
  - 1. Endocardial cushion
  - 2. Atrioventricular canal
  - 3. Conotruncal septation
  - 4. Atrial septum formation
    - a. Septum primum
    - b. Septum secundum
  - 5. Interventricular septum formation
- E. Six Aortic Arches
  - 1. Maxillary arteries branch from external carotid artery (ECA) - 1st pair
  - 2. Stapedial arteries - 2nd pair

3. Proximal common carotid artery and distal internal carotid artery (ICA) - 3rd pair
4. Left Ao arch - left 4th
5. Proximal right subclavian - partial component of the right 4th
6. Rudimentary not present in humans - 5th pair
7. Left pulmonary artery and ductus - left 6th pair
8. Right pulmonary artery - right 6th pair

F. Cardiac Valve Formation

1. The atrioventricular valves and supporting apparatus
2. The semilunar valves

G. Comparison of Fetal and Postnatal Circulation

1. Prenatal circulation
2. Umbilical vein
3. Ductus venosus
4. Eustachian valve
5. Foramen ovale
6. Ductus arteriosus

H. Neonatal Circulation

1. Ligamentum arteriosum
2. Fossa ovalis
3. Ligamentum venosum
4. Ligamentum teres

Section VII: 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 marker" refers to the orientation ridge on the transducer. Indicator mark is used with a clock reference for each view.
   b. The term "vertex up or vertex down" refers to the ultrasound image on the display monitor. When imaging congenital heart disease in the pediatric echocardiography laboratory, images are usually oriented in the "anatomically 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. Shaddy RE, Penny DJ, Feltes TF Cetta, F, Mital S. Moss and Adams' Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult, 10th Edition. Lippincott, Williams & Wilkins. Philadelphia, Pennsylvania. 2021
   b. Lai WW, Mertens LL, Cohen MS, Geva T. Echocardiography in pediatric and congenital heart disease: From Fetus to Adult, 3rd edition. John Wiley & Sons Inc, Hoboken. New Jersey. 2022

VII: 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
   b. Right atrial appendage is broad-based, triangular-shaped
2. Left atrial morphology
   a. Receives pulmonary veins
   b. Smooth-walled
   c. Left atrial appendage is thin, narrow-based shaped, pectinate muscles within chamber
3. Common atrium
   1. Absent septum secundum, septum primum
   2. Associated with isomerism of the atrial appendages (heterotaxy syndrome)
   3. 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

7. 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

b. Ventricular segment {D, L, X} – second letter of segmental set

i) D – Loop; morphologic right ventricle on the right side

ii) L – Loop; morphologic left ventricle on the right side

iii) X – Unable to determine looping

c. Great artery segment {S, I, D, L, A} – third letter of segmental set

i) S – solitus; normal position of the aortic valve, rightward and posterior to the pulmonic valve

ii) I – inversus; aortic valve is leftward and posterior to the pulmonic valve

iii) D – d-malposed; aortic valve is rightward and anterior to the pulmonic valve

iv) L – l-malposed; aortic valve is leftward and anterior to the pulmonic valve

v) A – anterior; aortic valve directly anterior to the pulmonic valve

8. Relational anatomy and cardiac position

a. Abdominal and viscero-atrial situs

a. Liver

b. Inferior vena cava

c. Descending aorta

b. Cardiac location

i) Levoposition

ii) Mesoposition

iii) Dextroposition

iv) Ectopia cordis

c. Cardiac orientation

i) Levocardia

ii) Mesocardia

iii) Dextrocardia

d. Segmental alignment and connections

i) Atrioventricular concordance versus discordance

ii) Ventriculoarterial concordance versus discordance

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

e. Assessment of cardiac structural morphology

i) Determine direction where apex is pointing with reference to axis of the heart

ii) Display all structures in correct anatomical position

9. Imaging technique

a. Subcostal

i) Coronal view - situs view (vertex up or down); transverse cut

- Indicator marker at 3 o'clock
  - 2D + color clip: cross-section of inferior vena cava, descending aorta, spine, liver, stomach
  - 2D + color sweep: hepatic veins to inferior vena cava to right atrium; continue to sweep to demonstrate cardiac apex

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

- Indicator marker at 12 o'clock (vertex up); 6 o'clock (vertex down)
  - 2D + color clip: inferior vena cava
  - 2D + color clip: abdominal descending aorta
  - PW: abdominal descending aorta below diaphragm

iii) Coronal view - long-axis view (vertex down)

- Indicator marker at 3 o'clock; posterior to anterior sweep
  - 2D + color sweep: inferior vena cava to right ventricular outflow tract
  - Focused 2D + color sweep at low Nyquist limit: atrial septum

iv) Left anterior oblique view

- Indicator marker between 4-5 o'clock; posterior to anterior sweep
  - 2D + color sweep: atrial septum
  - 2D + color clip: tricuspid valve and mitral valve en face

v) Sagittal view- short-axis view (vertex down)

- Indicator maker at 6 o'clock, right to left sweep
  - 2D + color sweep: bicaval view to left ventricle apex
  - Focused 2D + color sweep at low Nyquist limit: atrial septum
  - 2D + color clip: right upper pulmonary vein
  - 2D + color clip (+/- PW, CW): right ventricular outflow tract

vi) Right anterior oblique view

- Indicator marker between 1-2 o'clock
  - 2D + color clip: right ventricle inflow and outflow

b. Apical views (vertex down)

i) 4-Chamber

- Indicator marker at 3 o'clock; posterior to anterior sweep
  - 2D + color sweep: coronary sinus to right ventricular outflow tract
  - 2D + color sweep at low Nyquist limit: ventricular septum
  - Focused 2D clip: right atrium and right ventricle (right ventricle-centered view)
  - 2D + color clip: tricuspid valve
  - PW + CW (+/-) tissue Doppler: tricuspid valve
  - CW: tricuspid regurgitation jet (if present)
  - 2D + color clip (+/- PW, CW): right ventricular outflow tract
  - Focused 2D clip: left atrium and left ventricle
  - Color + PW: one right pulmonary vein
  - 2D + color clip: mitral valve
  - PW + CW + tissue Doppler: mitral valve
  - 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

ii) 2-Chamber

- Indicator marker at 2 o'clock
  - 2D clip: left atrium and left ventricle
  - 2D + color clip: mitral valve
  - Measurements: left atrial end-systolic area; left ventricular end-diastolic and end-systolic area and length

iii) 3-Chamber

- Indicator marker at 11 o'clock
  - 2D + color clip: left ventricular outflow tract, aortic valve, and proximal aorta
  - PW + CW: left ventricular outflow tract, aortic valve, and proximal aorta

c. Parasternal views (vertex up)

i) Parasternal long axis

- Indicator marker at 10 o'clock; sweep inferiorly towards right ventricular inflow, then sweep anteriorly towards right ventricular outflow tract
  - 2D + color sweep: reference to tricuspid valve, back to reference to pulmonic valve
  - Focused color sweep: ventricular septum
  - 2D + color clip: mitral valve
  - 2D + color clip: aortic valve
  - 2D + color clip: tricuspid valve

- CW: tricuspid regurgitation jet, if present
- 2D + color clip: pulmonic valve
- PW + CW: PV
- 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

ii) Parasternal short axis

- Indicator marker at 2 o'clock; sweep from cardiac base to apex
  - 2D + color sweep: aortic valve to apex then back to aortic valve to main and branch pulmonary arteries
  - Focused color sweep: ventricular septum
  - 2D + color clip: aortic valve
  - Focused simultaneous 2D + color clip (color-compare): left main, left anterior descending (LAD), and left circumflex (LCX) coronary arteries
  - Focused simultaneous 2D + color clip (color-compare): right coronary artery
  - 2D + color clip: mitral valve
  - 2D + color clip: tricuspid valve
  - CW: tricuspid regurgitation, if present
  - 2D + color clip: pulmonic valve
  - PW + CW: pulmonic valve, main pulmonary artery, proximal branch pulmonary arteries
  - 2D clip and M-mode: LV cross-section at level of papillary muscles
  - 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

iii) High left parasternal sagittal (ductal) view (vertex up)

- Indicator marker at 12 o'clock
  - 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
  - PW + CW: patent ductus arteriosus, if present

d. Suprasternal notch views

i) Suprasternal short axis (vertex up)

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

- 2D + color sweep: ascending aorta superiorly to first branch bifurcation, showing arch descending to the right or left during sweep with color mapping
- 2D + color sweep: leftward aspect of left innominate vein (exclude left superior vena cava or partial anomalous pulmonary venous return to left innominate vein)
- 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)

ii) Suprasternal long axis (vertex up)

- Indicator marker at 12 o'clock; sweep leftward towards left pulmonary artery
  - 2D + color clip: aortic arch and its branches
  - PW + CW: distal transverse aortic arch, aortic isthmus, and proximal descending aorta
  - Measurements: proximal and distal transverse aortic arch and aortic isthmus diameters

iii) Right parasternal sagittal view (vertex up)

- Indicator marker at 12 o'clock; right side of sternum
  - 2D + color clip: superior vena cava, inferior vena cava, atrial septum (exclude sinus venosus defect)

iv) Right parasternal transverse view (vertex up)

- Indicator marker at 3 o'clock; right side of sternum
  - 2D + color clip: right upper pulmonary vein draining into left atrium below level of right pulmonary artery

e. Calculations

i) Left ventricular end-diastolic and end-systolic endocardial volumes

ii) Left ventricular end-diastolic epicardial volume

iii) Left ventricular mass

iv) Left ventricular mass-to-volume ratio

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

vi) Left ventricular M-mode shortening fraction

vii) Left atrial volume

viii) Right ventricular systolic pressure from tricuspid regurgitation gradient or from ventricular septal defect gradient

ix) Pulmonary artery systolic pressure from patent ductus arteriosus gradient

10. Z-scores

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

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

c. 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

d. 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

B. Congenital Heart Disease

1. Anomalies of venous return

a. Anomalies of systemic venous return

i) Left superior vena cava draining into right atrium via coronary sinus

ii) Left superior vena cava draining into left atrium

iii) Retroaortic innominate vein

iv) Levoatrial cardinal vein

v) Right superior vena cava to left atrium

vi) Superior vena cava aneurysm

vii) Interruption of the inferior vena cava

viii) Absence of hepatic segment with azygous vein continuation

ix) Absence of the abdominal segment with hemiazygous continuation

x) Inferior vena cava draining into left atrium

xi) Absence of right superior vena cava

xii) 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

xiii) Hepatic veins drain into left atrium

xiv) Common atrium

b. Etiology

i) Prevalence

- Isolated anomalies occur in 0.3-0.5% of general population
- Occurs in 2-10% of patients with other forms of congenital heart disease
- Occurs in >70% of patients with heterotaxy syndrome
  - Abnormalities in regression of umbilical, vitelline, or cardinal venous segments during embryologic development
  - Abnormalities of visceroatrial development; bilateral “sidedness” (heterotaxy) syndrome

c. Pathophysiology

i) Normal physiology of systemic venous return to the right atrium

- Left superior vena cava to an intact coronary sinus with intact right superior vena cava
- Left superior vena cava to an intact coronary sinus with atresia of the right superior vena cava
- Interrupted inferior vena cava with azygous continuation
- Absent right superior vena cava with left superior vena cava to coronary sinus
- Abnormal physiology with systemic and pulmonary venous mixing

- Left superior vena cava draining into left atrium
- Right superior vena cava draining into left atrium
- Left superior vena cava to an “unroofed” coronary sinus
- Inferior vena cava draining into left atrium

d. Clinical presentation

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

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

e. Associated lesions

i) Coarctation of the aorta

ii) Mitral valve abnormalities, including congenital mitral stenosis

iii) Heterotaxy syndrome with polyploid

iv) Hypoplastic left heart syndrome

f. Echocardiographic evaluation

i) Subcostal coronal (transverse) and sagittal view of the abdominal viscera to image hepatic veins, inferior vena cava

ii) Subcostal sagittal view “caval” view to assess superior and inferior vena cava

iii) Suprasternal and high right/left parasternal views (vertex up) to image superior venous anatomy

- 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

g. 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

h. Echocardiographic assessment postoperatively

i) Evaluation of patency of repaired venous structures

ii) Evaluation for baffle leaks with baffle procedures

iii) Assessment of ligated venous structures

2. Anomalies of pulmonary venous return

a. Types

i) Normal pulmonary venous connections with abnormal drainage

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

ii) Partial anomalous pulmonary venous return (PAPVR)

- Drainage of one or more pulmonary veins anomalously to the systemic venous system or right atrium with normal drainage of remaining veins to the left atrium

- Scimitar syndrome - right pulmonary veins drain infracardiac to the hepatic system or inferior vena cava

iii) Total anomalous pulmonary venous return (TAPVR)

- Supracardiac
  - Pulmonary venous drainage into a vertical vein, coursing superiorly to innominate vein, draining to the superior vena cava
- Cardiac
  - Pulmonary venous drainage to the posterior aspect of the right atrium
  - Pulmonary venous drainage into the coronary sinus
- Infradiaphragmatic
  - Pulmonary venous drainage into a vertical vein, coursing inferiorly to portal venous system, ductus venosus, hepatic veins, or inferior vena cava
- Mixed type
  - Combination of anomalous sites of pulmonary venous return

b. Etiology

i) Incidence

- Partial anomalous pulmonary venous return: up to 25% of population may have a single right or left upper vein draining anomalously
- Total anomalous pulmonary venous return: 9 in 100,000 of population
  - Occurs mostly sporadically
  - Failure of embryologic incorporation of one or more pulmonary veins to the LA
  - Partial anomalous pulmonary venous return can be associated with Turner and Noonan syndromes

ii) Total anomalous pulmonary venous return can be associated with Allagille, Trisomy 22 (Cat-eye), Holt-Oram, and polysplenia/asplenia syndromes

c. Pathophysiology

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

ii) Pulmonary over circulation and congestion

iii) Pulmonary hypertension (PHTN)

d. Clinical presentation (pediatric and adult patients)

i) Tachypnea/dyspnea

ii) Cyanosis

iii) Congestive heart failure associated with supracardiac total anomalous pulmonary venous return

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

v) “Scimitar” sign on x-ray (scimitar syndrome)

e. Associated lesions

i) Hypoplastic left heart syndrome

ii) Heterotaxy syndrome with asplenia

iii) Coarctation of the aorta

f. Echocardiographic assessment (pediatric and adult patients)

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

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

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

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

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

vi) Total anomalous pulmonary venous return - assess confluence of pulmonary veins posterior to the LA

vii) Identify individual veins and their course

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

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

x) Treatment (surgical repair)

- Supracardiac and infradiaphragmatic type total anomalous pulmonary venous return - direct anastomosis of pulmonary vein confluence to left atrium
- Cardiac (coronary sinus) type total anomalous pulmonary venous return - unroofing of coronary sinus with closure of interatrial communication
- Scimitar syndrome partial anomalous pulmonary venous return - intra atrial baffle of right pulmonary veins to left atrium

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

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

ii) Evaluate for residual vertical vein

iii) Evaluate for pulmonary venous baffle obstruction

iv) TEE may be indicated for diagnostic quality imaging in adult patients

3. Anomalies of the atria

a. Atrial septal defect (ASD)

b. Types

i) Patent foramen ovale (PFO)

ii) Secundum (ASD)

iii) Primum (ASD)

iv) Sinus venosus defect (SVD)

v) Coronary sinus defect

c. 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

d. Patent foramen ovale

i) Overlapping of septum primum and septum secundum

ii) Located in the fossa ovalis region

e. Secundum atrial septal defect; mostly sporadic causes

i) Most common adult congenital heart 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

f. Primum atrial septal defect

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

- Sinus venosus defect

iii) Superior vena cava type sinus venosus defect

- Approximately 8% of all types of ASDs
- Sinus venosus not properly incorporated into the right atrium
- Deficient wall between the superior vena cava and right upper pulmonary vein
- Superior vena cava overrides right and left atrium
- Located in superior and posterior portion of atrial septum
- Associated with partial anomalous pulmonary venous return

g. Inferior vena cava 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

h. Coronary sinus (CS-ASD) 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 atrioventricularis of membranous septum and inferior vena cava orifice

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

i. 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)

j. 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

k. 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

vii) May have wide fixed-split second heart sound

l. Associated lesions

i) Secundum atrial septal defect and patent foramen ovale

- Mitral valve prolapse (MVP)
- Tricuspid atresia
- Hypoplastic left heart syndrome
- Total anomalous pulmonary venous return
- Ebstein's anomaly

ii) Primum atrial septal defect

- Atrioventricular septal defect
- Cleft mitral valve
- Atrioventricular valve regurgitation

iii) Sinus venosus defect

- Anomalous pulmonary venous return

m. 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

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

o. May need agitated saline (microcavitation) imaging

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

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

p. Transesophageal echocardiography (TEE) may be indicated

i) Evaluate type of defect

ii) Monitor intraprocedural progress and guidance for device closure

q. 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

r. 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

4. 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 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

ii) Left superior vena cava 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

- Apical 4-chamber view (vertex down) to assess membrane
  - Doppler interrogation of fenestration gradient (if present)

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

ii) Treatment

- No treatment necessary if membrane non-obstructive
- Surgical resection of obstructive membrane with atrial septal defect closure

iii) Echocardiographic assessment postoperatively (pediatric and adult patients)

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

5. Atrioventricular valve anomalies

a. Atrioventricular septal defect (AVSD)

i) Etiology

- Abnormal development of endocardial cushions
- 0.19:1000 live births
- 4.0-5.0% of all congenital heart defects (CHD)
- 40% of children with trisomy 21 and congenital heart disease have an atrioventricular septal defect
- Failure of septum primum to fuse with endocardial cushions

ii) Categorized as complete, partial (incomplete), or transitional (intermediate)

- Complete atrioventricular septal defect consists of primum atrial septal defect, common atrioventricular valve, inlet ventricular septal defect
    - Balanced
    - Unbalanced
        - ~ Left ventricular dominant
        - ~ Right ventricular dominant
    - Partial (incomplete) type atrioventricular septal defect consists of primum atrial septal defect, common atrium, cleft mitral valve
    - Transitional (intermediate) type atrioventricular septal defect consists of two complete atrioventricular valve orifices, primum atrial septal defect, restrictive ventricular septal defect

iii) Pathophysiology

- Defect between the two atria and/or ventricles based on type of atrioventricular septal defect
- Incomplete type same as atrial septal defect, complete type same as ventricular septal defect (VSD)
- Volume overload
- Pressure overload

iv) Clinical presentation (pediatric and adult patients)

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

v) Echocardiographic evaluation (pediatric and adult patients)

- Subcostal coronal, sagittal views (vertex down) to assess atrial septal defect(s), atrioventricular valve(s), ventricular septal defect(s)
- 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)
- 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
- 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
- Parasternal short axis (vertex up) to assess atrioventricular valve morphology, papillary muscle size and location
  - Assess for cleft, double orifice

vi) Echocardiographic findings should define:

- Type of defect (complete, transitional/intermediate, partial/incomplete)
- Size of ventricles (balanced or unbalanced)
- Degree of atrioventricular valve regurgitation

vii) Assess for signs of pulmonary hypertension

- Ventricular septal defect gradient
- Right atrioventricular valve regurgitation jet

viii) Related testing

- 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
- Chest x-ray

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

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

ix) Treatment (pediatric and adult patients)

- Surgical patch repair of atrial septal defect(s) and ventricular septal defect, if present
  - Single-patch technique
- Suture repair of "cleft" in anterior leaflet of left atrioventricular valve
- Replacement of left or right atrioventricular valve if significant stenosis or regurgitation is unrepairable
- Resection or relief of subaortic obstruction, if present

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

- Evaluate for residual shunt
- Evaluate for atrioventricular valve stenosis and regurgitation
- Evaluate for signs of pulmonary hypertension
- Evaluate for aortic valve regurgitation
- Evaluate for left ventricular outflow tract obstruction
- Evaluate ventricular function

b. Ebstein anomaly

i) Etiology

- 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

ii) Pathophysiology

- Varying degrees of tricuspid regurgitation
- Systemic venous congestion with hepatomegaly

iii) Clinical presentation (adult and pediatric patients)

- Mild forms may be asymptomatic (present in adolescence or adulthood)
- Severe forms (presenting as a fetus or neonate)
  - Cyanosis
  - Cardiomegaly (wall-to-wall heart) on x-ray
  - Tachypnea
- Split first and second heart sound

- Arrhythmias - supraventricular tachycardia (SVT)

iv) Associated lesions

- Patent foramen ovale or secundum atrial septal defect, 30-78%
- Pulmonary valve stenosis
- Pulmonary valve atresia or "functional" atresia

v) Echocardiographic evaluation (pediatric and adult patients)

- 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)
- Subcostal left anterior oblique view (vertex down) to assess enface view of tricuspid valve
  - Allows visualization of leaflet components, and chordal attachments
- Subcostal right anterior oblique view (vertex down)
  - Allows visualization of the tricuspid valve, chordal attachments into the right ventricular outflow tract, and pulmonic valve
- Apical 4-chamber view (vertex down) to assess right atrium and right ventricle, determine portion of "atrialized" right ventricle, visualize the degree of tricuspid valve, apical displacement
  - Perform Celermajer index
- 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
- Echocardiographic findings may include paradoxical ventricular septal motion, large right atrium (depending on the severity of apical displacement), assess for signs of pulmonary hypertension

vi) Related testing

- ECG
  - Tall P waves in limb lead II
  - Right bundle branch block (RBBB)
  - 30% will have Wolff-Parkinson-White syndrome
- Chest x-ray
  - Cardiomegaly with prominent right-sided border
  - Decreased pulmonary blood flow

vii) Treatment

- No treatment necessary in mild cases
- Severe cases, an initial palliation, Blalock-Thomas-Taussig shunt (BTTS)
- Surgical repair (Cone, Danielson, Carpentier procedures)
- Valve repair/replacement

viii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate for residual tricuspid valve insufficiency and/or iatrogenic tricuspid valve stenosis
- Evaluate for residual atrial/ventricular level shunt, if history of atrial septal defect/ventricular septal defect repair
- Evaluate right ventricular outflow tract, pulmonic valve, and branch pulmonary arteries
- Evaluate ventricular function
- Evaluate for signs of pulmonary hypertension

c. Tricuspid valve atresia

i) Etiology

- 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
- 0.039-0.085:1000 live births, 2.7% of all congenital heart disease
- Initial survival is dependent upon interatrial communication
- May occur with normally related great vessels or transposition of great vessels
- May occur with a normal pulmonary artery and large ventricular septal defect or pulmonary stenosis and small ventricular septal defect or pulmonary atresia. by adulthood will have gone through surgical palliation(s)
    - Fontan procedure

ii) Pathophysiology

- Atresia of the tricuspid valve
    - Systemic venous return travels though atrial communication - patent foramen ovale or atrial septal defect to left atrium
    - Desaturation due to systemic venous return mixing with pulmonary venous return to left atrium
- Ventriculo-arterial relationships and the size of the ventricular septal defect will largely determine clinical presentation
    - Restrictive ventricular septal defect will result in subaortic stenosis in d-transposition of the great arteries
- Ventricular volume overloading; dependent on amount of pulmonary flood flow
    - Ventricular enlargement increases ventricular wall stress, which results in compensatory hypertrophy
    - Hypertrophy – impaired diastolic function
    - Subendocardial ischemia
- 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

iii) Clinical presentation

- Signs and symptoms
  - Cyanosis at birth
  - Tachypnea
  - Poor feeding

iv) Associated lesions

ix) Patent foramen ovale, atrial septal defect

x) D-loop transposition of great arteries

xi) Left superior vena cava to coronary sinus

xii) Ventricular septal defect

xiii) Pulmonary valve stenosis

xiv) Pulmonary atresia

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

xvi) Juxtaposition of the atrial appendages

v) Echocardiographic evaluation

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

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

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

xx) 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

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

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

vi) Treatment

xxiii) Staged palliation

- First stage
  - ~ Unrestricted pulmonary outflow- pulmonary artery banding
  - ~ Severe pulmonary valve stenosis or pulmonary atresia: modified Blalock-Thomas-Taussig shunt
  - ~ 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)

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

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

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

xxvi) Evaluate ventricular function

d. Hypoplastic left heart syndrome

i) Etiology

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

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

xxix) Often associated with coarctation of aorta

xxx) 0.103 - 0.267:1000 live births

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

ii) Pathophysiology

- Postnatal survival depends upon patency of the ductus arteriosus, which supplies the systemic circulation
  - o "Ductal-dependent" lesion; ductus arteriosus patency must be maintained (PGE1 infusion, ductal stenting)
- Surgical palliation performed within the first week of life (stage 1 Norwood procedure)
- Unobstructed interatrial communication must be maintained
- Staged palliation resulting in total cavopulmonary anastomosis

iii) Clinical presentation

- Pediatric patients in the neonatal period
  - o Varying degrees of cyanosis at birth
  - o At 3-5 days post birth, develop profound metabolic acidosis due to closing ductus
  - o Tachypnea
  - o Shock
- Palliated patients with complications (infant to adult)
  - o Evidence of congestive heart failure (right ventricular dysfunction/severe TR)
  - o Tachypnea
  - o Diminished distal pulses with upper/lower extremity pressure differential (aortic re-coarctation)
  - o Pleural effusions
  - o Ascites (Fontan patients with protein-losing enteropathy)

iv) Echocardiographic features (pediatric and adult patients)

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

v) Echocardiographic evaluation (newborn/unpalliated patients)

- Subcostal coronal, left anterior oblique views (vertex down) to assess size and flow through patent foramen ovale/atrial septal defect
- Subcostal sagittal views (vertex down) to assess abdominal aortic flow, systemic venous return, ventricular function
- 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
- Parasternal long (vertex up) to assess right atrium, right ventricle, and tricuspid valve, assess left side structures for size and patency
- Parasternal short axis (vertex up) evaluate coronary artery origins, patent ductus arteriosus, right ventricular function
- Suprasternal short axis, high left parasternal view (vertex) to assess branch pulmonary arteries, and patent ductus arteriosus
- Suprasternal long axis (vertex up) to assess aortic arch and patent ductus arteriosus

vi) Treatment

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

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate patency of shunts/conduits (stage 1)
- Evaluate neoaortic arch for obstruction
- Evaluate interatrial communication patency (must be widely patent)
- Evaluate for tricuspid regurgitation
- Evaluate ventricular function

- Evaluate systemic inflow and outflow for obstruction
- Evaluate cavopulmonary anastomoses/conduits to ensure patency

6. Ventricular anomalies
   a. Ventricular septal defect (VSD)
      i) Etiologies
         xxxii) Perimembranous
            - Most common type of ventricular septal defect
            - 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

         xxxiii) 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

         xxxiv) Trabecular (muscular)
            - Excessive cavitation of myocardial tissue during formation of ventricular septum and ventricular walls

         xxxv) 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

         xxxvi) 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
               ~ Associated with muscular aortic stenosis and/or aortic arch anomalies (interrupted aortic arch)

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

xxxviii) Increased pulmonic flow

xxxix) Left atrial enlargement

xl) 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

iii) Clinical presentation (pediatric and adult patients)

xli) 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
- Large defects (unrestrictive)
  - ~ Cardiomegaly on x-ray
  - ~ Congestive heart failure
  - ~ Low frequency pansystolic murmur at lower left sternal edge or no systolic murmur
- Pulmonary hypertension (late, with large-volume shunts)
  - ~ Eisenmenger's syndrome

iv) Echocardiographic features (pediatric and adult patients)

xlii) Defect in ventricular septum

xliii) Left heart enlargement with large, unrestrictive defects

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

xlv) Increased pulmonic flow

xlvi) Evaluate RV size and function

xlvii) Evaluate for PHTN

xlviii) Estimate RV pressure by measuring gradient

xlix) Enlarged LA and LV

v) Echocardiographic views

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

li) 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

lii) 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

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

liv) 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

vi) Treatment

lv) Small (restrictive) ventricular septal defects

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

lvi) 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 should not be closed

vii) Echocardiographic assessment postoperatively

lvii) Evaluate for residual shunt

lviii) Evaluate for additional (undetected) ventricular septal defects

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

lx) Evaluate for pulmonary hypertension

lxi) Assess outflow tract obstruction

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

i) Etiology

lxii) 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

lxiii) Associated lesions

- Perimembranous ventricular septal defect
- Ebstein anomaly of the morphologic (systemic) ventricle
- Right ventricular outflow tract obstruction

- Congenital complete heart block

ii) Pathophysiology

- Normal hemodynamic physiology
- Morphologic right ventricle - systemic cardiac output
- Morphologic left ventricle - pulmonary cardiac output
- Right ventricle remodels to accommodate systemic pressure
- Left ventricle atrophies under pulmonary pressure
- Ebsteinoid tricuspid valve can lead to malcoaptation and regurgitation
- Severe volume load on right ventricle can cause congestive heart failure in infancy
- Patients with complete heart block have higher mortality
- Right ventricular dysfunction may occur later in life

iii) Clinical presentation (pediatric and adult patients)

- May be clinically silent until 3rd or 4th decade of life if no associated congenital heart disease
- Congestive heart failure caused by tricuspid regurgitation and/or systemic (right) ventricular failure
- Development of complete heart block

iv) Echocardiographic features (pediatric and adult patients)

- Apical 4-chamber view demonstrates attachment of left-sided atrioventricular valve (tricuspid) valve slightly apical to right-sided atrioventricular valve (mitral) valve
- Parasternal short axis view demonstrates leftward and anterior displacement of the aorta (L-transposition of the great vessels)
- Morphologic RV is the systemic ventricle
    - To evaluate RV function, dp/dt and Tei index may help

v) Echocardiographic views

- This lesion can be confusing; it is helpful to draw a picture of the patient's anatomy, to use as a guide while scanning
- Subcostal coronal and left anterior oblique views (vertex down) to assess for atrial level defect, left ventricular outflow tract obstruction, and ventricular septal defects
- Subcostal sagittal views (vertex down) to assess ventricular function
- Apical 4-chamber view (vertex down) to assess and confirm ventricular arrangement, cardiac function, tricuspid valve morphology, function and competency, assess outflow tracts
- Parasternal long axis view (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
- Parasternal short axis view (vertex up) to confirm ventricular morphology, evaluate ventricular sizes and systolic function, assess for ventricular septal defect(s)

vi) Treatment

- No treatment necessary for asymptomatic cases with no additional congenital heart disease
- Moderate to large unrestrictive ventricular septal defect - primary closure
- Relief of pulmonary/subpulmonary stenosis, if present
- Patients with Ebsteinoid tricuspid valve and significant tricuspid regurgitation - pulmonary artery banding
- Complete heart block - pacemaker insertion
- "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

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- 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
- Evaluate systemic ventricular (right ventricle) function (all patients)
- Assess presence and degree of tricuspid regurgitation
- Evaluate for residual ventricular septal defect - status post repair
- Assess arterial connections for obstruction (patients with double-switch operation)
- Assess atrial baffles for obstruction/leaks (patients with double switch operation)

7. Semilunar valve anomalies

a. Aortic stenosis (AS)

i) Types and etiology

- Subvalvular tunnel
  - Hypoplastic left ventricular outflow tract
- Left ventricular outflow tract obstruction associated with hypertrophic cardiomyopathy (HCM)
  - Systolic anterior motion of mitral valve chordae and apparatus causing left ventricular outflow tract obstruction
  - May be isolated mid-cavitary
- Discrete subaortic membrane
  - Associated with
    - ~ Ventricular septal defect or spontaneously closed VSD
    - ~ Shone's complex
  - Fibrous or fibromuscular tissue in left ventricular outflow tract
    - ~ Circumferential or partial within LVOT

- Bicuspid aortic valve
  - Incidence 2% of population
  - Associated with coarctation of the aorta
  - Congenitally malformed leaflets
- Congenital aortic valve stenosis
  - Three leaflets
  - Varying degrees of commissural fusion
- Unicuspid aortic valve
  - Congenitally malformed leaflets
  - Single slit-like commissure in cross-section
- Quadricuspid aortic valve
  - Congenitally malformed leaflets
    - ~ Varying degrees of commissural fusion
  - Rare; Incidence 0.013%
- Supravalvular aortic stenosis
  - Narrowing of ascending aorta
  - Can occur sporadically or familial (defect in elastin gene on chromosome 7)
  - Associated with William's syndrome

ii) Pathophysiology

- Increased resistance to flow from left ventricle
- Concentric left ventricular hypertrophy (LVH) develops
  - Calculation of LV mass assists with evaluating the left ventricular workload
- Left atrial enlargement may also occur in patients with LVH
  - Associated with abnormal left ventricular diastolic function

iii) Clinical presentation

- 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)

iv) Echocardiographic features

- Thickened leaflets
- Doming of leaflets with restriction of excursion
- Concentric LVH
- LV diastolic function
    - May have diastolic “fluttering” of anterior MV leaflet if significant aortic insufficiency is present
- Doppler pressure gradient
- Ascending aortic dilation
- Evaluate for subvalvular stenosis
- Evaluate for supravalvular stenosis
    - Calculate Z-scores for the aortic annulus and root, sinotubular junction, and ascending aorta

v) Echocardiographic views

- Subcostal left anterior oblique views (vertex down) to assess left ventricular outflow tract, aortic valve, and proximal ascending aorta
- Apical 4-chamber view (vertex down) to assess cardiac function, presence of left ventricular hypertrophy
- Apical 5-chamber view (vertex down) to assess left ventricular outflow tract, aortic valve, and proximal ascending aorta
- Parasternal long axis view (vertex up) to assess left ventricular outflow tract, and aortic valve cusp appearance and mobility
- Parasternal short axis view (vertex up) evaluates aortic valve cusps and valve function, assess for left ventricular hypertrophy
- High right parasternal sagittal view to assess ascending aorta
    - View provides an optimal spectral Doppler angle
- Pedoff transducer (adolescents to adults)
    - Right parasternal window
    - Suprasternal notch window
    - Apical 5-chamber window

vi) Treatment

- Catheterization
    - Balloon dilation
- Surgery
    - Valve repair
    - Ross procedure
    - Valve replacement

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate transvalvular pressure gradient
- Evaluate for aortic valve regurgitation
- Evaluate for prosthesis stenosis/failure/paravalvular leak

- Evaluate left ventricular function
- Assess ascending aorta and arch
- Evaluate right ventricle to pulmonary artery connection and branch pulmonary arteries (Ross procedure)

b. Pulmonary stenosis

i) Types and etiology

- Pulmonary valve stenosis
  - Bicommissural pulmonary valve
  - Tri-leaflet valve with cusp fusion
- Hypoplastic pulmonary valve annulus and conus
  - Thickened domed and stenotic valve
  - Narrow or hypoplastic valve annulus
- Supravalvular
  - Discrete supravalvular membrane
  - "Hourglass" deformity in main pulmonary artery segment
  - Branch pulmonary artery stenosis
- Subvalvular pulmonary stenosis
  - Prominent muscle bundle at the infundibular opening, creating subvalvular obstruction

ii) Pathophysiology

- Increased resistance to flow from right ventricle
- Obstruction results in increased RV systolic pressure
- Afterload increase results in RV hypertrophy and muscle mass

iii) Clinical presentation

- May be asymptomatic
- Systolic murmur
- Critical pulmonary valve stenosis
  - Cyanosis

iv) Echocardiographic features

- Thickened leaflets
- Doming of leaflets
- RVH
- Doppler pressure gradient
- Evaluate for infundibular stenosis

v) Echocardiographic views

- 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

- 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
- Apical 4-chamber view with anterior sweep (vertex down) to assess cardiac function, right ventricular outflow tract and pulmonary valve
- Parasternal long axis view (vertex up) to assess right atrium, right ventricle, tricuspid valve, and pulmonic valve
- Parasternal short axis view (vertex up) evaluate right ventricular outflow tract, pulmonic valve, main and branch pulmonary arteries

vi) Treatment (pediatric and adult patients)

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

vii) Post-operative/repair evaluation

- Evaluate trans-valvular pressure gradient
- Evaluate for residual pulmonary valve stenosis, presence/severity of pulmonary valve regurgitation
- Assess for residual right ventricular outflow tract obstruction
- Evaluate right ventricular size and function
- Assess tricuspid valve regurgitation

c. Pulmonary atresia with intact ventricular septum

i) Etiology

- Environmental factors
- Flow disturbances through the right heart during embryologic and fetal development
- 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

ii) Pathophysiology

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

- Pulmonary blood flow is dependent on ductus arteriosus (ductal-dependent lesion)
- Suprasystemic right ventricular pressures with and without coronary sinusoids can result in myocardial ischemia
- Pulmonary artery flow is ductal dependent at birth unless major aorto-pulmonary collateral arteries (MAPCAs) are present (disease variant)

iii) Clinical presentation

- Cyanosis at birth
- May present with congestive heart failure, hepatomegaly in subtype with massive tricuspid regurgitation and right heart enlargement
- Adolescents and adults may present with congestive heart failure due to poor LV function and development of mitral valve regurgitation
- Myocardial infarction (MI) may occur due to presence of right ventricular dependent coronary circulation (RVDCC)

iv) Echocardiographic features

- Absent or diminished RVOT with no flow demonstrated from RV to PA
- Varying degrees of TV/RV hypoplasia usually present
- Coronary sinusoids/fistulae to the RV may be present
- PDA commonly originates from the underside of the arch with a tortuous path to pulmonary arteries
- Collaterals or MAPCAs are rare in this (PA/IVS) subtype

v) Echocardiographic views

- Subcostal coronal, left anterior oblique, and sagittal views to assess systemic venous return, atrial septal defect, right atrium/right ventricle size and function
- Subcostal right anterior oblique view (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
- Apical 4-chamber view (vertex down) to assess cardiac function, assess right ventricle for sinusoids, assess tricuspid valve
- Parasternal long axis view (vertex up) to assess right atrium, right ventricle, and tricuspid valve
- Parasternal short axis view (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
- Suprasternal short axis view, high left parasternal view (vertex up) to assess branch pulmonary arteries, and patent ductus arteriosus

vi) Treatment

- 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)

- Pulmonary valve perforation and balloon dilation possible in cases with adequate tricuspid valve and right ventricular size and function
- Heart transplantation may be necessary in cases with right ventricular dependent coronary circulation (RVDCC)

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Assess left ventricular function
- Evaluate atrial shunting
- Assess presence and severity of mitral regurgitation
- 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)

8. Aortic arch anomalies

a. Coarctation of the aorta

i) Types

- Pre-ductal (arch hypoplasia)
- Juxta-ductal (discrete “shelf”)
- Post-ductal and “mid-aortic”

ii) Etiology

- Luminal narrowing of the aorta from the normal diameter (usually isthmus)
- Flow disturbances through the left heart during embryologic and fetal development
- Environmental factors
- Occurs in 5-7% of all congenital heart disease
- 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)
- Syndromic associations
  - Turner syndrome
  - 22q11 deletion (DiGeorge syndrome)

iii) Pathophysiology

- Narrowing of aorta at level of coarctation
- Increased resistance to flow at the coarctation site with drop in pressure distally
- Usually see a difference in blood pressure between arms and legs
- Collateralization occurs late, decompressing proximal arch segment

iv) Clinical presentation

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

v) Echocardiographic views

- Subcostal sagittal view (vertex down or up) focused clip of abdominal aorta, spectral doppler performed at the level of the diaphragm
- Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess atrial level shunt, cardiac function
- Apical 4-chamber view (vertex down) to assess cardiac function
- Apical 5-chamber view (vertex down) to assess left ventricular outflow tract and flow velocities across aortic valve
- Parasternal long axis view (vertex up) to assess and measure the aortic valve annulus, sinuses, sinotubular junction, and ascending aorta
- Parasternal short axis view (vertex up) evaluate aortic valve morphology, cardiac function, presence of patent ductus arteriosus
- Suprasternal short axis view, to evaluate aortic arch sidedness, assess presence of patent ductus arteriosus, and evaluate the descending aorta
- Suprasternal long axis view, 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

vi) Treatment

- Surgery
    - End-to-end anastomosis (discrete narrowing)
    - Extended end-to-end anastomosis (diffuse hypoplasia)
- Catheterization
    - Balloon dilation with or without stent placement (adolescents and adults)

ii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate for recurrence or residual coarctation
- Assess for dilation or aneurysm formation (especially adults with history of patch augmentation)
- Assess ventricular function
- Evaluate for un-masked atrioventricular or mitral valve obstructive lesions

b. Interrupted aortic arch

i) Types

- Type A (30%); discontinuity between last vessel (usually left subclavian artery) and aortic isthmus
- Type B (70%); discontinuity between left carotid and left subclavian arteries
- Type C (<1%); discontinuity between carotid arteries or between the left carotid artery and the right innominate (brachiocephalic) artery
- All types may have aberrant left or right subclavian arteries

ii) Etiology

- Abnormal regression of arch segments during embryology

- Environmental factors may play role
- Highly associated with 22q11 deletion (DiGeorge syndrome)
- 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

iii) Pathophysiology

- Interruption of aorta with ductal dependent circulation to post-ductal organs
- Ascending aorta provides coronary and cerebral perfusion
- Posterior malalignment ventricular septal defect is most commonly present
- Ductal closure causes circulatory shock

iv) Clinical presentation

- Cyanosis as newborn
- Metabolic acidosis and circulatory shock due to ductal closure
- Ductal closure results in cessation of flow to abdominal aorta and abdominal organs
- Repaired patients may present with symptoms of re-coarctation

v) Echocardiographic views

- Subcostal coronal, left anterior oblique, and sagittal views (vertex down) to assess left ventricular outflow tract and proximal ascending aorta
- Apical 5-chamber view (vertex down) to assess left ventricular outflow tract, ventricular septal defect, and presence and degree of subaortic narrowing
- Parasternal long axis view (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
- Parasternal short axis view (vertex up) to evaluate aortic valve morphology, patent ductus arteriosus
- Suprasternal short axis view (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)
- Suprasternal long axis view (vertex up) to evaluate brachiocephalic vessels to determine type of interruption, assess aortic arch, and patent ductus arteriosus

vi) Treatment

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

- Ventricular septal defect closure
- Resection of obstructive subaortic conus tissue
- Ross-Konno repair for significant aortic annular hypoplasia
- Patients with severe subaortic stenosis and/or significant aortic valve hypoplasia may need Norwood-type arch reconstruction with Damus-Kaye-Stansel (DKS) and Rastelli-type ventricular septal defect baffle with right ventricle to pulmonary artery homograft

vii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate arch for residual obstruction
- Assess left ventricular outflow tract for obstruction
- Assess for residual ventricular septal defect(s)
- Evaluate ventricular function
- Assess pulmonary artery connection (homograft/conduit) in cases with Norwood/Rastelli type repairs

9. Conotruncal anomalies

a. Transposition of the great arteries

i) Types

- Dextro-transposition of the great arteries (D-TGA) – isolated ventriculo-arterial discordance
- Congenitally corrected transposition of the great arteries (CCTGA) – atrio-ventricular and ventriculo-arterial discordance; (covered in ventricular anomalies)

ii) Etiology of D-TGA

- Abnormal conotruncal septation resulting in ventriculo-arterial discordance
- Higher incidence in maternal women, infant of diabetic mother
- Environmental factors may play a role
- 10th most common congenital heart disease
- 2nd most common cyanotic lesion

iii) Pathophysiology of D-TGA

- Two parallel circuits exist
- Systemic venous blood returns to the right ventricle and then is recirculated to the systemic circulation through the aorta
- Pulmonic venous blood returns to the left ventricle and then is recirculated to the pulmonary circulation through the pulmonary artery

iv) Clinical presentation D-TGA

- Cyanotic at birth
- Single loud second heart sound
- Ventricular septal defects/left ventricular outflow tract obstruction variants will have flow murmur
- Coarctation variants may have significant metabolic acidosis

v) Associated lesions

- Ventricular septal defects in 40-45% of patients
- Left ventricular outflow tract obstruction in 20-30%
- Coarctation of the aorta
- Systemic venous anomalies

vi) Echocardiographic features

- Ventriculo-arterial discordance
- Fibrous continuity between the mitral and PV
- RV infundibulum is located between the aorta and the TV
- Coronary artery anatomy has a variable pattern
- Ventricular configuration appears normal in the neonatal period
- Systemic morphologic RV (Mustard and Senning patients) becomes enlarged with septal flattening

vii) Echocardiographic views

- 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
- Subcostal sagittal views (vertex down) will demonstrate a parallel relationship of the great arteries
- 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
- Parasternal long axis view (vertex up) will demonstrate the parallel relationship of the great arteries, assess left ventricular outflow tract, and ventricular septal defects
- Parasternal short axis view (vertex up) evaluate semilunar valve morphology, coronary artery origins and proximal courses, patent ductus arteriosus
- Suprasternal short axis view, high left parasternal view (vertex up) to assess branch pulmonary arteries, and patent ductus arteriosus
- Suprasternal long axis view (vertex up) to assess aortic arch and patent ductus arteriosus

viii) Common treatment

- Balloon atrial septostomy
  - Immediate palliation for extreme cyanosis; improves atrial level shunting
- 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

ix) Echocardiographic assessment postoperatively (pediatric and adult patients)

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

b. Tetralogy of Fallot (TOF)

i) Types

- TOF with pulmonary stenosis (varying degrees)
- TOF with pulmonary atresia
- TOF with absent PV syndrome
- TOF with complete atrioventricular septal defect (AVSD)

ii) Etiology

- Occurs in 4-8% of all congenital heart disease
- Embryologic maldevelopment of the subpulmonary conus
- Results in narrowing of the infundibulum and an anterior malalignment VSD
- Associated defects:
  - Right aortic arch
  - Vascular ring
  - Coronary artery anomalies
  - Systemic venous anomalies
  - Branch pulmonary artery discontinuity
- Syndromic associations:
  - 22q11 deletion (DiGeorge syndrome)
  - Holt-oram
  - Trisomy 13, 18, 21

- Alagille syndrome
- Goldenhar syndrome

iii) Pathophysiology

- Combination of obstructive pulmonary outflow and large ventricular septal defect result in ventricular-level right-to-left shunt and cyanosis
- Degree of cyanosis directly related to severity of pulmonary outflow obstruction
- Ductal patency improves pulmonary blood flow but may contribute to pulmonary overcirculation in cases with mild pulmonary outflow obstruction

iv) Clinical presentation

- Varying degrees of cyanosis
  - Significant pulmonary outflow obstruction increases ventricular right-to-left shunt, resulting in increased cyanosis
  - Cases of tetralogy of Fallot with absent pulmonary valve syndrome usually have massively dilated PA branches causing airway compression, exacerbating cyanosis
  - May present with over-circulation and signs of congestive heart failure ("pink TOF")
- Murmur
  - Harsh systolic murmur for typical cases
  - Harsh systolic and diastolic murmurs in TOF with absent PV
- Abnormal x-ray (boot-shaped heart)
- Repaired adult patients may present with signs and symptoms of right heart failure from pulmonary regurgitation or residual outflow obstruction
- Repaired adult patients frequently experience inter-ventricular conduction delays and ventricular dysrhythmia
- Unrepaired adolescent or adult patients present with profound cyanosis, polycythemia and fingertip clubbing

v) Echocardiographic features

- Short axis view; cases of TOF with absent PV may have right heart enlargement due to significant associated pulmonary regurgitation
- Abnormalities of aortic arch sidedness and branching may be present

vi) Echocardiographic views

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

- 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
- Parasternal long axis view (vertex up) to assess classic features of TOF, aortic override of large anterior malalignment ventricular septal defect, and right ventricular hypertrophy (RVH)
- Parasternal short axis view, high parasternal views (vertex up) demonstrates 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
- Suprasternal short axis view (vertex up), evaluate aortic arch sidedness, and branching pattern
- Suprasternal long axis view (vertex up) to assess aortic arch and patent ductus arteriosus

vii) Treatment

- Repair dependent on anatomy
    - May need staged approach if anatomy unfavorable for repair
        - ~ Blalock-Taussig-Thomas shunt early
        - ~ Complete repair involving ventricular septal defect closure and relief of pulmonary stenosis (around) 6-months of age
- Transannular patch repair
- Non-transannular repair
- Complete repair with right ventricle to pulmonary artery (RV-PA) homograft/conduit
- Complete repair with plication of main pulmonary artery and branch pulmonary arteries (TOF, absent pulmonary valve)
- 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 (Melody, Sapien)

viii) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate aorta-to-pulmonary shunt
- Evaluate for residual ventricular septal defects
- Assess right ventricular outflow tract for residual obstruction and possible aneurysm formation
- Evaluate pulmonic valve for residual stenosis or regurgitation
- Evaluate pulmonary arteries for residual narrowing or dilation
- Evaluate tricuspid valve regurgitation
- Assess ventricular function

- Particular attention to right ventricular function

c. Double outlet right ventricle

i) Types

- 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

ii) Etiology

- Maldevelopment of conotruncus resulting in both great arteries arising predominantly or entirely from the RV
- Occurs in 1-2% of all patients with congenital heart disease (CHD)
- Associated syndromes (12%)
  - Trisomy 13, 18
  - 22q11 deletion (DiGeorge syndrome)
- Associated defects
  - Subpulmonary or subaortic outflow tract obstruction
  - Straddling atrioventricular valves (AVV)
  - Atrioventricular septal defect
  - Aortic arch anomalies
    - ~ Coarctation
    - ~ Arch branching malformations
- Systemic venous anomalies

iii) Pathophysiology

- 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

iv) Clinical presentation

- Newborn patients
  - Murmur
  - Varying degrees of cyanosis
- Adolescent and adult patients post repair
  - Palliated patients present with cyanosis
  - Right heart failure
  - Ventricular dysrhythmia
  - Congestive heart failure

v) Echocardiographic features

- All cases have malposed great artery relationship
- One artery may override VSD
- Subaortic or subpulmonary outflow obstruction
- Frequently muscular discontinuity between atrioventricular valves and semilunar valves (bilateral conus)

vi) Echocardiographic views

- 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
- Subcostal sagittal views (vertex down) will demonstrate a parallel relationship of the great arteries
- 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
- Parasternal long axis view (vertex up) will demonstrate the parallel relationship of the great arteries, assess outflow tracts, and ventricular septal defect(s)
- Parasternal short axis view (vertex up) evaluate great artery relationship
- Suprasternal short axis view, high left parasternal view (vertex up) to assess aortic arch sidedness, branch pulmonary arteries, and patent ductus arteriosus
- Suprasternal long axis view (vertex up) to assess aortic arch and patent ductus arteriosus

vii) Treatment

- 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

- Staged single ventricle palliation for cases with:
  - Straddling atrioventricular valves
  - Remote ventricular septal defect
  - Hypoplastic ventricles

viii) Echocardiographic assessment postoperative

- Evaluate great artery anastomosis sites for obstruction
- Evaluate for residual ventricular septal defect
- Assess outflow tracts for residual obstruction
- Evaluate pulmonary arteries
- Evaluate for tricuspid valve regurgitation
- Assess ventricular function

Section VIII: 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 qualification

VIII: 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
6. Wall motion abnormalities
   a. Hypokinesis
   b. Akinesis
   c. Dyskinesis
   d. Hyperkinesis
   e. Wall motion scoring index
7. Global LV systolic performance
   a. Ejection phase indices

i) Ejection fraction (EF)
ii) Fractional shortening (FS)
iii) Velocity of circumferential fiber shortening (Vcf)
iv) Cardiac output (CO) and SV
v) Tissue Doppler imaging (TDI)
vi) Speckle tracking echocardiography (STE)

b. Non-ejection phase indices
i) Systolic time intervals
- Pre-ejection time

ii) LV dp/dt
iii) Acceleration time of aortic flow
iv) Pressure/volume analysis

c. Indirect methods
i) Mitral E-end point septal separation (EPSS)
ii) Aortic root motion
iii) Descent of cardiac base
iv) Sphericity Index of LV
v) Velocity time integrals (VTI) of outflow
- LVOT
- RVOT

vi) Potential limitations
- Endocardial dropout
- Influence of load conditions
- Regional wall motion abnormalities
- Some methods require no major shape distortion
- Oblique measurements/angles
- Limited frame rate of 2D imaging
- LV foreshortening
- Translation artifact
- Desynchrony of contraction

vii) Improve endocardial delineation
- Harmonic imaging
- Contrast agents (enhanced cardiac ultrasound)

8. Assessment of ventricular systolic function
a. Fractional shortening
i) Technique
- M-mode

- 2D

ii) Calculations

iii) Normal values

iv) Limitations

- Less accurate in the presence of wall motion abnormalities
- Load dependent

b. Stroke volume (SV)

c. Cardiac output and index

d. Ejection fraction

i) Characteristics

- Ratio of SV to DV
- Most commonly used clinical index of LV function
- Influenced by preload, afterload and heart rate
- Visual estimate valid with interobserver variability

ii) Calculations

iii) Normal values

iv) Limitations

- Due to altered loading conditions
  - Mitral regurgitation: % ejection fraction normal with intrinsic myocardial dysfunction
  - AS: % ejection fraction decreased with normal myocardial function
  - Severe anemia
  - Hemodialysis patients

e. Left ventricular mass

i) M-mode method

- Penn formula
- ASE formula
- Normal values

ii) 2D method

- Area-length
- Truncated ellipsoid
- Normal values
- Limitations

f. Change in pressure by change in time (dP/dt)

i) Characteristics

- Relatively load-independent

ii) Calculations

iii) Appropriate measurement placement

iv) Normal values

g. LV volumes

i) Simpson's rule and modified Simpson's rule

- Biplane more accurate
- Single plane
  - Suitable if LV symmetric
- Quick method (regression equation)

ii) End systolic volumes (ESV)

- Most reproducible volume measure
- Insensitive to cardiac loading
- Powerful predictor of cardiac events
- Normal values

iii) End diastolic volumes (EDV)

- Endocardium more difficult to image at end diastole
- More variable than end systolic volumes
- Normal values

iv) Technical considerations

- Image optimization
- Both atrioventricular values imaged
- Avoid aorta and coronary sinus
- Selection of precise time in cardiac cycle for measurement
- ED frame with largest LV cavity just after MV closure
- ES frame with smallest LV cavity just before MV opening
- Enhanced cardiac imaging

h. Tissue Doppler imaging

i) Characteristics

ii) Appropriate technique

9. LV quantification

a. Prognosis

i) Coronary artery disease (CAD)

ii) Acute MI

iii) Cardiomyopathy

- Volume/mass ratio

b. Methods for regional function

i) 17 segment wall score

ii) Tissue Spectral Doppler imaging

- Strain

c. Considerations
   i) Hypertension
      - LV mass predicts morbidity/mortality
      - LV wall thickness/cavity dimension predicts morbidity/mortality
   ii) Interdependence of LV and RV
   iii) Global RV systolic function

d. Associated conditions
   i) LVH
   ii) Ischemic heart disease
   iii) Dilated cardiomyopathy (DCM)
   iv) Hypertrophic cardiomyopathy (HCM)
   v) Restrictive cardiomyopathy (RCM)
   vi) RV pressure and volume overloads
   vii) Valvular heart disease
   viii) Pericardial disease
   ix) Transplant rejection
   x) Takotsubo cardiomyopathy
   xi) Chemotherapy cardiotoxicity

e. Therapeutic implications
   i) Angiotensin-converting enzyme inhibitors
   ii) Timing of surgery in volume overload states (MR,AI)
   iii) Timing of surgery in pressure overload states (AS)

10. Assessment of ventricular diastolic function

a. General principles
   i) Four phases of diastole
      - Isovolumic relaxation
      - Early rapid diastolic filling
      - Diastasis
      - Late diastolic filling caused by atrial contraction
   ii) Parameters of diastolic function
      - Relaxation
        - Isovolumic relaxation time
        - Maximum rate of pressure decline (dP/dt)
        - Time constant of relaxation (tau)
      - Compliance
        - dP/dV
        - Chamber stiffness constant
      - Ventricular diastolic pressures

- Ventricular diastolic filling curves

b. Echocardiography evaluation

i) 2D imaging

- Chamber dimensions
- Wall thickness
- Automated endocardial border tracing

ii) Doppler imaging

- Assessment of MV inflow & normal values
  - E-wave/A-wave ratio
  - Deceleration time (DT)
  - Isovolumic relaxation time (IVRT)
  - Mitral E-wave velocity (E)
  - Mitral A-wave velocity (A)
  - Mitral A-wave duration
  - Time from MV opening to e-wave velocity
- 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 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)

c. LV filling patterns

i) Technical factors

- Sample volume location
- Doppler modality (PW versus CW)
- Intercept angle

ii) Physiologic factors

- Respiration
- Heart rate
- PR interval
- Age
- Preload

- Afterload
- Exercise
- Valsalva
- LV systolic function
- Atrial contraction function
- Diastolic function
- Cardiac function

iii) Normal Doppler values

- Aortic valve regurgitation (AR)
- Aortic valve regurgitation duration – A duration
- Deceleration Time (DT)
- E/A
- e’
- Isovolumic relaxation time (IVRT)
- Vp using color M-mode

iv) Abnormal relaxation

- Doppler values
  - Aortic valve regurgitation
  - Aortic valve regurgitation duration < mitral a duration
  - Deceleration time
  - E/A
  - e’
  - Isovolumic relaxation time
  - Vp
- Significance
  - Initial stage
  - No increase in mean LA pressure
  - Difficult to assess with elevated heart rate
  - Seen in
    - ~ Systemic hypertension
    - ~ Coronary artery disease
    - ~ Cardiomyopathies
    - ~ Elderly

v) Pseudonormalization

- Doppler values
  - Aortic valve regurgitation
  - Aortic valve regurgitation duration < mitral a duration

- Deceleration time
- E/A
- e’
- Isovolumic relaxation time
- S/D
- Vp

- Significance
  - Spectral Doppler of mitral inflow appears normal
  - Underlying relaxation abnormally with elevated LA pressure
  - Preload reduction may unmask relaxation abnormality
    - ~ Valsalva
    - ~ Nitroglycerin
  - Seen in
    - ~ Dilated cardiomyopathy
    - ~ Restrictive cardiomyopathy
    - ~ Ischemic cardiomyopathy

vi) Restrictive/noncompliance

- Doppler values
  - Aortic valve regurgitation
  - Aortic valve regurgitation duration < mitral a duration
  - Deceleration time
  - E/A
  - e’
  - Isovolumic relaxation time
  - S/D
  - Vp
- Significance
  - Late stage
  - Severe decrease of LV compliance
  - Elevated mean LA pressure at rest
  - May reverse with preload reduction maneuvers

vii) Estimation of LV filling pressures

- Normal LV filling pressures
  - Abnormal relaxation pattern except with severe LVH
- Elevated LV filling pressures

d. RV quantification

i) Dimensions and volumes

- Basal, mid and base-apex dimensions

- Systolic and diastolic volumes
- RV wall thickness

ii) Systolic function

- Tricuspid annular plane systolic excursion (TAPSE)
- Tricuspid annular s' velocity
- Fractional area change

iii) Other quantitative assessment

- Pulmonary vascular resistance
- RV index of myocardial performance

iv) RV diastolic function

- Tricuspid inflow E/A
- e', a', and s' velocity
- E/e' ratio

Section IX: Cardiomyopathies

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

IX: CARDIOMYOPATHIES

A. Hypertrophic Cardiomyopathy

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

- Anterior papillary muscle displacement and elongation
- Other situations
  - Mitral valve repair
  - Atrioventricular replacement
  - Hypovolemia
  - Endogenous and exogenous catecholamines
- Elongated anterior mitral leaflet (AML)
- Increased area of AML
- Calcified mitral annulus

e. Left atrial enlargement

f. Narrowed left ventricular outflow tract diameter

g. Altered texture of myocardium

h. Basal septal endocardial thickening (contact lesion)

7. M-mode features

a. Normal or small left ventricular size

b. Extent of hypertrophy

c. Normal or increased shortening fraction

d. Left atrial enlargement

e. Abnormal mitral valve motion

i) Reduction of E-F slope

ii) Systolic anterior motion of leaflet

iii) Mitral annular calcification (MAC)

f. B-bump

g. AV mid-systolic closure

h. Color M-mode of mitral valve and left ventricular outflow tract obstruction

8. Doppler features

a. Diastolic dysfunction assessment

i) Characteristics

- Impaired relaxation
- Decreased compliance
- Increased left ventricular end diastolic pressure
- Decreased EA ratio
  - Normal filling pattern
    - ~ Normalization by mitral regurgitation
    - ~ Normalization by increased left atrial pressure
- Poor correlation between deceleration time and left atrial pressure
- 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

iv) Primary MR versus secondary MR

c. Level of obstruction (LVOT versus midcavity)

i) Doppler wave patterns

- Late peaking
- Late systolic

ii) Provocative maneuvers

d. Limitations and pitfalls

i) Differential diagnosis

- Chronic hypertension
- Cardiac amyloid
- Pheochromocytoma
- Freidreich's ataxia
- Inferior MI with previous LVH
- Dynamic LVOT obstruction not specific
- Hyperdynamic LV systolic function
  - Anemia
  - Volume depletion
  - Catecholamines
- Hypertensive hypertrophic cardiomyopathy in elderly
- Postoperative period
- Sub aortic obstruction post AVR
- Acute anteroapical MI with pre-existing basal septal hypertrophy

e. Related testing

i) ECG

ii) Laboratory testing

iii) Correlative imaging

f. Common treatments

i) Role of echo

- Evaluation of therapy
- Pacemaker implementation and therapy
- Septal ablation
- Myotomy/myectomy

B. Dilated Cardiomyopathy

1. Epidemiology
    a. Prevalence
    b. Genetics
2. Etiologies
3. Clinical presentations
    a. Signs and symptoms
    b. Physical findings
    c. Auscultatory finding
4. 2D features
    a. Chamber enlargement
    b. Global hypokinesis of LV
    c. Regional wall motion abnormality
    d. Mural thrombi in apex
    e. Aneurysm
    f. Spontaneous echo contrast
5. M-mode features
    a. Chamber dilation
    b. Hypokinesis of LV
    c. Decreased LV wall thickening
    d. Decreased fractional shortening
    e. Increased end point septal separation
    f. Reduced aortic root motion
    g. Reduced ejection fraction
6. Doppler features
    a. MR and TR assessment
    b. Diastolic dysfunction assessment
    c. RV systolic pressure assessment
    d. LVEDP assessment
7. Prognostic role of echo
    a. EF
    b. DT
    c. RV function
8. Related testing
    a. ECG
    b. Laboratory testing
    c. Correlative imaging
9. Common treatments

C. Restrictive cardiomyopathy

1. Etiologies of RCM
    a. Primary
        i) Idiopathic
    b. Secondary
        i) Amyloid heart disease
        ii) Hemochromatosis
        iii) Heart muscle disease
            - Post-irradiation heart disease
            - Carcinoid heart disease
            - Doxorubicin/daunorubicin toxicity
            - Progressive systemic sclerosis
        iv) Eosinophilic endomyocardial disease
2. Classifications of Infiltrative CM
    a. Infiltrative
        i) Interstitial
            - Amyloid
            - Hemochromatosis
            - Sarcoid
            - Malignancy
        ii) Storage
            - Glycogen
            - Lipid
3. Clinical presentations
    a. Signs and symptoms
    b. Physical findings
    c. Auscultatory finding
4. 2D features
    a. Diffuse speckling or granular appearance of myocardium (amyloid)
    b. Pericardial or pleural effusion
    c. Bi-atrial enlargement
    d. Small to normal LV cavity
    e. Possible LVH
    f. Normal systolic function
5. M-mode features
    a. Normal LV size
    b. Concentric LVH
    c. Decreased LV shortening fraction
    d. Pericardial or pleural effusion

6. Doppler features
    a. MR and TR assessment
    b. Diastolic dysfunction assessment including Tissue Doppler imaging
    c. Minimal or absent respiratory variation in Doppler flows
7. Differential diagnosis
    a. Constructive pericarditis
8. Related testing
    a. ECG
    b. Laboratory testing
    c. Correlative imaging
9. Common treatment

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

Section X: Cardiac Tumors/Masses

1. List benign and malignant cardiac tumors
2. Describe echocardiographic criteria used for evaluating tumors

X: CARDIAC TUMORS/MASSES

- A. Thrombus
  - 1. Echocardiographic characterization
    - a. Location and site of attachment
    - b. Size
    - c. Degree of mobility
    - d. Shape
    - e. Co-existing cardiac abnormalities
  - 2. Potential for embolization
    - a. Size
    - b. Protrusion into cavity
    - c. Mobility
  - 3. Technical suggestions
    - a. Use high-frequency, short-focus transducers
    - b. Decrease depth of field
    - c. Move focal zone nearer apex
    - d. Use nonstandard views
    - e. Contrast enhanced echocardiography
  - 4. Pitfalls
    - a. Artifact
    - b. Must differentiate from prominent trabeculations, papillary muscles, anomalous chords
- B. Benign
  - 1. Rhabdomyoma
  - 2. Myxoma
  - 3. Pericardial teratoma
  - 4. Fibroma
  - 5. Hemangioma
  - 6. Papillary fibroelastoma
- C. Malignant
  - 1. Sarcomas
  - 2. Malignant lymphoma
  - 3. Miscellaneous others
  - 4. Characteristics
    - a. Etiology

b. Common locations

5. Echocardiographic features

a. Occur more frequently in right heart

b. Sessile, with intracavitary extension

D. Secondary (Metastatic)

1. Most often metastasize to pericardium with associated effusion

2. Occur more frequently in right heart

3. Tumors invading right heart from inferior vena cava

a. Wilms tumor (nephroblastoma)

4. Characteristics

a. Etiology

b. Clinical presentations

i) Signs and symptoms

ii) Physical findings

iii) Auscultatory findings

E. 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

F. Differentiation from Other “Masses”

1. Extracardiac

2. Intracardiac

a. Moderator band

b. Papillary muscles

c. Redundant chordae

d. Myxomatous tissue

e. Vegetations

f. Tuberculomas

g. Chiari network
h. Eustachian valves
i. Catheter or pacemaker wire
j. Others

3. Artifacts
   a. Side lobes
   b. Reverberation
   c. Improper gain settings

Section XI: Coronary Artery Anomalies

1. Describe the normal anatomy and function of coronary arteries
2. Identify common and rare types of coronary artery anomalies, and explain their clinical significance and underlying causes
3. Recognize the signs and symptoms associated with coronary artery abnormalities
4. Identify the sonographic findings of coronary artery anomalies
5. Discuss management strategies and treatment options for coronary artery anomalies, including when intervention is warranted
6. Relate coronary artery anomalies to other congenital heart diseases

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XI: CORONARY ARTERY ANOMALIES

- A. Coronary Artery Anatomy and Function
  - 1. Coronary arteries
    - a. Flow distribution
    - b. Artery location, ostia, proximal common stem, termination
    - c. Relation between coronary arteries and myocardial wall segments
    - d. Flow measurements and limitations
    - e. Coronary flow reserve
    - f. Myocardial perfusion with contrast-enhanced echo
- B. Normal Coronary Artery Mapping
  - 1. PSAX is the best view for proximal origin
    - a. Coronary arteries originate above the level of the aortic valve and therefore will not be visualized in the same plane as the aortic leaflets
  - 2. Visualizing the coronary arteries require superficial anterior views and will not be in the standard planes of imaging
  - 3. Technical
    - a. High frequency transducer
    - b. Lower compression
    - c. Lower 2D gain
    - d. Optimizing depth settings
    - e. Color low scale and increase gain
- C. Right Coronary Artery
  - 1. PLAX can visualize the most proximal take off above the sinotubular junction
    - a. Most parallel to flow
  - 2. Mid portion is visualized angled towards the TV
  - 3. Distal portion
    - a. Right ventricular inflow tract (RVIT) view distal RCA in posterior right AV groove in ventricular sulcus
  - 4. PSAX with a slight counterclockwise turn
    - a. RCA at right aortic sinus
    - b. 11 o’clock position
    - c. Proximal RCA with a portion of the mid-RCA slight counterclockwise rotation of

transducer with anterior angulation

d. Apical – visualized in the A4 and A5 views for the take off

i) Distal RCA is also seen with a posterior angulation in the AV groove

e. Right sternal border

i) Transducer marker at noon

ii) Patient in right decubitus position provides a better view of the proximal and mid-RCA

D. Left coronary system

1. PLAX look between the aorta and PA
2. PLAX view angling anterolaterally toward the RVOT and lateral left ventricular wall define the more distal LAD and LCX
3. PSAX with a slight clockwise turn slight leftward and anterior angulation
4. Left main at left posterior sinus
   a. 4 o'clock position
   b. Proximal LCX will also be seen
5. Apical views
   a. Anterior angulation of the apical long-axis/three-chamber view; the distal LCX can be visualized in the AV groove
   b. Anterior apical four-chamber view with focus on the apex; distal segments of the LAD can be imaged in this view

E. Coronary Artery Abnormalities

1. Anomalies can occur in the form of fistulas, anomalous origins, congenital coronary aneurysm, or myocardial bridging
   a. These anomalies could affect function
   b. Some congenital coronary anomalies are associated with cardiac arrest and sudden death
2. Anomalies include
   a. Anomalous origins from the pulmonary artery
   b. Anomalous aortic origin from opposite sinus of the aortic root (AAORCA, AAOLCA)
   c. Myocardial bridging
   d. Congenital coronary aneurysm
   e. Coronary fistula
3. Hemodynamically significant anomalies include
   a. Anomalous origins from the pulmonary artery ALCAPA
   b. Deep myocardial bridging
   c. Congenital fistula
   d. ALCAPA – Anomalous left main coronary artery originates from the PA left or posterior sinus
4. Etiology
   a. 1:300,000 live births

b. May occur alone or associated with VSD, PDA, AVSD, transposition of the great arteries (TGA), etc.

5. Left coronary artery
   a. Neonatal presentation
      i) LCA empties into the pulmonary artery (steal)
      ii) Lower pulmonary artery pressures cause blood to course backward in the left main and left anterior descending coronary arteries and enter into the pulmonary artery
   b. Late (childhood to adult) presentation
      i) Collaterals develop for myocardial perfusion
      ii) RCA provides blood flow to the collaterals
   c. Clinical signs and symptoms
      i) Neonate-failure to thrive, diaphoresis, tachypnea, chest pain, congestive heart failure, sudden cardiac death
      ii) Adolescence/teenage murmur (usually from mitral regurgitation), shortness of breath, congestive heart failure
   d. Echo findings
      i) Left coronary artery from the pulmonary artery
      ii) Retrograde flow in left coronary artery
      iii) Abnormal flow in pulmonary artery
      iv) Dilated right coronary artery from collaterals
      v) Depressed left ventricular systolic function
      vi) Mitral regurgitation to varying degrees result in left atrial dilation
   e. Other coronaries arising from the pulmonary artery include right coronary artery, left anterior descending or circumflex
      i) Patients with anomalous right coronary artery from anomalies may be asymptomatic
      ii) Left anterior descending and circumflex origins from the pulmonary artery may show ischemia
      iii) If all three coronary arteries arise from the pulmonary artery, the infant shows profound heart failure
   f. Treatment
      i) The left coronary artery may be reimplanted into the aorta
      ii) A tunnel may be created to direct the left coronary artery to the aorta
6. Left circumflex artery from the right aortic sinus of Valsalva
   a. Most common functional artery anomaly is the left circumflex artery from the right aortic sinus
   b. Origin of the circumflex from the aorta and separate from the right coronary may be seen
7. Left main coronary artery from the right aortic sinus of Valsalva
   a. Origin of the left main from the right sinus of Valsalva
   b. May take different routes

i) Anterior to the pulmonary artery
ii) Posterior to the aorta
iii) In the interventricular septum
iv) Between the aorta and pulmonary artery

c. Intramural course of the coronary artery
i) Coronary artery course between the two great vessels

d. Associated with sudden cardiac death

e. Clinical presentation
i) Patient has syncope episodes, presyncope, and chest pain associated with exercise
ii) Patients may be asymptomatic
iii) Stress test may be normal or show myocardial ischemia

8. Right coronary artery from the left aortic sinus of Valsalva
a. Intramural course of the coronary artery
b. Right coronary artery course between the two great vessels
c. Associated with sudden cardiac death

9. Single coronary artery
a. One coronary ostium from one of the aortic sinuses
i) Follows a peripheral course and distribution of one or both coronary arteries

b. Majority of cases are asymptomatic
i) May be associated with sudden cardiac death
ii) Atherosclerotic occlusion of the single artery must be monitored

c. Clinical signs and symptoms
i) Relatively asymptomatic
ii) Chest pain
iii) Sudden cardiac death (exercise induced)

d. Echo findings
i) Myocardial ischemia
ii) Mitral valve insufficiency to varying degrees results in left atrial dilation

e. Treatment
i) Reserved for patients with myocardial ischemia
ii) Surgical repair includes
- Bypass graft
- Reimplantation to the correct location

10. Myocardial bridging
a. Congenital anomaly where the artery is encased by myocardium
i) As the myocardium contracts, the bridge can squeeze the coronary artery
ii) Typically, the LAD is affected

b. Two types
    i) Superficial bridging
    ii) Deep bridging (>2mm deep)
c. Clinical presentation
    i) Usually benign if superficial
    ii) May also cause ischemia, infarcts, stunning or cardiac death
    iii) Symptoms present during exercise
        - Fatigue
        - Chest pain
        - Shortness of breath
d. Echo findings
    i) May or may not have regional wall motion abnormalities
e. Treatment
    i) Bypass graft
    ii) Remove the bridge that is causing symptoms

11. Congenital coronary fistulae
    a. Connection between a cardiac chamber and coronary artery
        i) Origin from the RCA
            - Empty into right ventricle
            - Empty into the left atrium or the right atrium
        ii) Origin from the LAD
            - Empty into the left ventricle
            - Empty into the left or right atrium
            - Usually see one site of termination
    b. Hemodynamics
        i) When shunt leads into right ventricle, hemodynamics may resemble a left to right shunt
        ii) When shunt is in left ventricle, flow may be similar to aortic insufficiency
        iii) Small and medium fistulas are only treated with symptoms
        iv) Large should be closed even if symptomatic
        v) There is a potential for a coronary artery steal
        vi) Usually see one site of termination
        vii) Fistulas can increase in size over time
        viii) Serial monitoring is essential
    c. Clinical signs and symptoms
        i) Asymptomatic
        ii) Chest pain
        iii) Congestive heart failure

iv) Continuous murmur

d. Associated congenital heart disease

i) Pulmonary atresia with an intact ventricular septum

e. Echo findings

i) Identified by continuous color Doppler flow

ii) Dilated coronary arteries

iii) Dilated chambers depending on site of entry of fistula

f. Treatment

i) Interventional devices

ii) Surgical closure

12. Associated congenital heart disease for coronary artery anomalies include:

a. Dextro-transposition of the great arteries

b. Truncus arteriosus

c. Tetralogy of Fallot

d. Levo-transposition of the great arteries (L-TGA)

e. Double inlet left ventricle

f. Single ventricle

g. Double outlet right ventricle

h. Pulmonary atresia with intact ventricular septum

i. Hypoplastic left heart syndrome

13. Post operation echo interrogation

a. Left ventricular function

b. Mitral valve regurgitation

c. Left atrial dilatation

d. Aortic valve insufficiency

e. Unroofing

14. Other imaging modalities

a. CMR

b. CT

c. Nuclear myocardial perfusion

d. Angiography

Section XII: Pericardial Disease

1. Identify the normal structure and function of the pericardium
2. Recognize etiologies and clinical presentations of pericardial effusion
3. Interpret echocardiographic features of pericardial disease
4. Distinguish between constrictive pericarditis, cardiac tamponade, and other pericardial disorders
5. Describe diagnostic and treatment approaches for pericardial diseases
6. Understand congenital pericardial anomalies and their complications

XII: PERICARDIAL DISEASE

A. Normal Pericardium

1. Structure
2. Function

B. Pericardial Effusion

1. Abnormal accumulation of fluid in the pericardial cavity
2. Etiologies
   a. Pericardial inflammation
   b. Infection
   c. Radiation
   d. Autoimmune diseases
   e. Trauma
   f. Neoplasms
   g. Iatrogenic
   h. Connective tissue diseases
   i. Metabolic diseases
   j. Chest trauma
   k. Aortic dissections
   l. Myocarditis
   m. Post MI
   n. Medications
3. Clinical presentations
   a. Signs and symptoms
      i) May have no symptoms at all
      ii) Shortness of breath
      iii) Chest pain
      iv) Painful breathing
      v) Low grade fever
      vi) Feelings of anxiousness
      vii) Rapid heart rate
      viii) Fainting/dizziness
      ix) Edema
   b. Physical findings

i) Quiet, hypodynamic heart

ii) Elevated jugular venous pressure (JVP)

c. Auscultatory finding

i) Distant heart sounds

ii) Ewart's syndrome

4. Types

a. Circumferential

b. Loculated

5. Size

a. Small 1cm; seen only in diastole

b. Moderate 1 to 2cm; seen in diastole and systole

c. Large > 2cm in width may result in swinging heart

6. Echocardiographic features

a. Anechoic space

b. Fluid is posterior to parasternal long and anterior to descending aorta

c. Variable size and location

7. Differentiation from pleural effusion, epicardial adipose, pericardial fat and other posterior echo-free spaces

8. Treatment

a. Medical

b. Pericardiocentesis

c. Pericardial window

d. Pericardiectomy

C. Constructive Pericarditis

1. Etiology

a. Idiopathic

b. Infectious

c. Immune inflammatory

d. Neoplastic disease

e. Post radiation

f. Post cardiac surgery

g. Other

2. Clinical presentations

a. Signs and symptoms

i) Dyspnea

ii) Positional 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) Diastolic pericardial knock

ii) Holosystolic functional regurgitation

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/M-mode features

a. Thickened pericardium

b. Lack of pericardial slide

c. Interventricular septal bulge to the left during inspiration

d. Normal LV size and EF

e. Enlarged atria

f. Flattening of LV posterior wall in diastole

g. Abrupt posterior motion of ventricular septum in early diastole (septal bounce)

h. Dilated inferior vena cava and hepatic veins

i. Ascites

5. Doppler features

a. Respiratory variation in velocities

i) Left inflow >25%

ii) Right inflow >40%

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

iv) TR velocity increased

d. Pulmonic outflow increases
e. Aortic outflow decreases
f. Hepatic vein increase in S and D forward flows
g. Expiration (reciprocal changes)
    i) Increased mitral E' velocity
    ii) Increased pulmonary venous diastolic forward flow
    iii) Decreased tricuspid E' velocity
    iv) Decreased loss of diastolic filling with marked expiratory reversal
h. Tissue Doppler
    i) Septal e velocity relatively normal or accentuated in constriction (> 12cm/sec)
    ii) Lateral wall tethering and e' is less than septal
    iii) Annulus reversos
i. Hepatic vein reversal in expiration
g. 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/Em 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 flow not typically restrictive
        iii) Hepatic vein flow not characteristics of constriction
7. Pitfalls
    a. High filling pressure
    b. Sample volume placement (translational motion)
    c. Atrial fibrillation
    d. Localized constriction
    e. Treatment
        i) Pericardiectomy

D. Cardiac tamponade
1. Etiologies
    a. Pericarditis
    b. Iatrogenic

c. Malignancy
d. Idiopathic
e. Infectious
f. Immune inflammatory
g. Collagen diseases
h. Trauma
i. Aortic dissection
j. Radiation
k. Post MI
l. Uremia
m. Other

2. Clinical presentation
   a. Signs and symptoms
      i) Chest pain
      ii) Dyspnea
   b. Physical findings
      i) Tachycardia
      ii) Hypotension
      iii) Narrow pulse pressure
      iv) Pulsus paradoxus
      v) Elevated (JVP)
      vi) Beck's triad
      vii) Ewart's sign
3. Auscultatory findings
   a. Diminished heart sounds
   b. Pericardial friction rub
4. ECG findings
   a. Electrical alternans
5. Correlative imaging
   a. Chest x-ray
   b. CMR
   c. CT
6. Hemodynamics
   a. Accumulation of pericardial fluid causes pericardial pressure to rise and compromise systemic venous return in RA
   b. Ventricular filling is impaired and cardiac output is decreased
   c. Intrapericardial pressure is influenced by both volume of fluid and rate at which it accumulates
   d. Dissociation of intrathoracic and intracardiac pressures due to insulating effect of

effusion

e. Relatively fixed combined cardiac volume (ventricular interdependence)

7. Echocardiographic features

a. 2D features

i) Pericardial effusion

ii) RA or RV early diastolic chamber collapse

iii) IVC plethora

iv) Dilated hepatic and SVC

v) Reciprocal changes in ventricular volumes (septal shift)

vi) “Swinging heart” if large effusion

vii) Inspiratory septal bounce of interventricular septum

b. Doppler features

i) Respiratory variation in velocities

- Left inflow >30%
- Right inflow >60%

ii) Inspiration

- Decreased mitral E’ velocity
- Decreased pulmonary venous diastolic forward flow
- Increased tricuspid E’ velocity
- Pulmonic outflow increases
- Aortic outflow decreases
- Hepatic vein increase in S and D forward flows

iii) Expiration (reciprocal changes)

- Increased mitral E’ velocity
- Increased pulmonary venous diastolic forward flow
- Decreased tricuspid E’ velocity
- Pulmonic outflow decreases
- Aortic outflow increases
- Decrease or loss of hepatic vein diastolic filling with marked expiratory reversal

iv) Technical caveats

- Use of respirometer preferred
- Typical respiratory patterns are reversed in a mechanically ventilated patient
- Changes may be seen in patients with high filling pressures, ASD or RVH

c. Surgical

i) Pericardial window

ii) Total/limited pericardiectomy

iii) Echo-guided pericardiocentesis

d. 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

e. Pigtail catheter introduced and left in for several days with intermittent drainage

f. Low complications rate

E. Other Pericardial Disease

1. Pericardial cyst

a. Characteristics

b. Echocardiographic features

i) Echolucent space

ii) Frequently adjacent to RA

c. Additional imaging is often needed

i) CT

ii) CMR

2. Congenital absence of pericardium

a. General

i) Uncommon

ii) More frequent in males

iii) Congenital or caused by surgery

iv) 30% will have other congenital abnormalities

b. Types

i) Total absence

ii) Partial absence

c. Echocardiographic features

i) Unusual echo windows

ii) Cardiac hypermobility

iii) Marked shift of cardiac chambers to the right

iv) Abnormal ventricular septal motion

v) Leads to enlargement of the RV

- Best seen in PLAX

d. Complications

i) Herniation or strangulation of LAA with partial absence

Section XIII: Acquired and Systemic Disease in the Pediatric Patient

1. Identify and describe the causes and echocardiographic features of acquired and systemic cardiac diseases in pediatric patients
2. Recognize the clinical presentations and complications of ineffective endocarditis and Kawasaki disease
3. Differentiate between various cardiac masses and structural abnormalities on echocardiogram
4. Understand the cardiac effects and imaging findings related to trauma, cardiotoxic therapies, and rheumatic heart disease in children
5. Apply appropriate imaging techniques for diagnosis and follow-up of pediatric cardiac conditions

XIII: ACQUIRED AND SYSTEMIC DISEASE IN THE PEDIATRIC PATIENT

A. Cardiac Trauma-Blunt Chest Trauma – Cardiac Injury Evaluation

1. Pericardial effusion
2. Pneumothorax
3. Ventricular septal defect
4. Aneurysm of right ventricular outflow tract
5. Pseudoaneurysm
6. Ruptured septal coronary
7. Valve rupture
8. Coronary arterial rupture/dissection
9. Chamber rupture

B. Ineffective Endocarditis: Microbial Infection of the Endothelial Lining of the Heart

1. Most common bacterial organisms
   a. Streptococcus viridans
   b. Staphylococcus aureus
2. At risk patients
   a. Ventricular septal defect
   b. Patent ductus arteriosus
   c. Aortic valve abnormalities
   d. Mitral valve abnormalities
   e. Tetralogy of Fallot
   f. Trauma to the endocardium from stress forces
   g. Indwelling catheters
   h. Intracardiac devices and prostheses
   i. Immunodeficiency
   j. Intravenous drug use
   k. Corrective and palliative surgeries
3. Pathophysiology/natural history
   a. Occurs primarily on cardiac valves, but may occur on other endocardial surfaces or intracardiac devices
   b. High mortality/complication rate
4. Clinical presentation
   a. Signs and symptoms

i) Fever
ii) Septicemia
iii) Fatigue/weakness
iv) “Flu-like” symptoms
v) Weight loss

5. Echo features: echogenic intracardiac mass
   a. Typically irregular shaped
   b. Pedunculated or sessile
   c. Rarely impairs valve motion
   d. Often flutter or vibrate
   e. Common locations
      i) Atrial side of mitral valve and tricuspid valve
      ii) Ventricular side of aortic valve and pulmonary valve
      iii) Located in right ventricle side for ventricular septal defect
      iv) Main pulmonary artery for patent ductus arteriosus
      v) May also occur in conduits, patch repairs and prosthetic valves
   f. Less common locations
      i) Chordae
      ii) Mural endocardium
      iii) Eustachian valve
      iv) Pacemaker wire
   g. New valvular regurgitation
      i) Secondary effects of valvular regurgitation
         - Abscess
         - New partial dehiscence of prosthetic valve
6. Complications
   a. Cusp or leaflet rupture/flail
   b. Perforation
   c. Abscess
   d. Aneurysm
   e. Fistulae
   f. Dehiscence of prosthetic valve
   g. Pericardial effusion
   h. Conduction disturbance
   i. Hemodynamic compromise
      i) Regurgitation (acute or chronic)
      ii) Valvular stenosis
      iii) Shunt

iv) Congestive heart failure

j. Embolization

i) Cerebral

ii) Systemic

iii) Pulmonary

7. 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. Calcified deposits

g. Myxomatous or flail mitral valve leaflet

C. Kawasaki Disease

1. General

a. Kawasaki is acute vasculitis syndrome that affects the coronary arteries

b. Inflammation of mid-sized arteries

c. Diagnosed by its principal signs and symptoms

d. Usually seen <5 years of age, affects males more than females

e. Winter – spring predominance

f. Exact pathogen not yet identified

2. Echocardiographic features

a. Myocarditis, pericarditis and coronary artery dilation

b. Pericardial effusion

c. Ventricular dysfunction

d. New valvular regurgitation

e. Coronary artery ectasia and/or dilation and aneurysm

f. Thrombus within coronary arteries

g. Late-stage coronary narrowing

3. Imaging coronaries

a. High frequency transducer

b. Optimizing depth settings

c. Lower 2D gain

d. Decreased color Doppler scale

e. Customized machine presets specific to vendor

f. Lower compression

g. Flow optimization

h. Write priority

D. Cardiotoxicity

1. Radiation therapy: type of cancer treatment that uses beams of intense energy to kill cells. Destroys the genetic material that controls how cells grow and divide the goal is to destroy as few normal, healthy cells as possible.
    a. Cardiac manifestation
        i) Diffuse fibrosis of the myocardium
        ii) Dense collogen and fibrin replace the normal adipose tissue of the outer layer of the heart leading to
            - Myocardial cell death
            - Effusion
            - Ischemia
            - Possible tamponade
            - Fibrosis
            - Pericardial fibrosis
            - Fibrotic changes to leaflets and valves; changes to valves on the left side are more common than those on the right
            - Myocardial fibrosis
    b. Echo interrogation
        i) Ventricular systolic and diastolic function
        ii) Valvular disease
        iii) Tissue velocity imaging
        iv) Strain-rate and strain analysis
    c. Chronic findings
    d. Pericardial disease, coronary artery disease, cardiomyopathy, valvular disease and conduction abnormalities can manifest years or decades after the original treatment
2. Chemotherapy is a drug treatment that uses powerful chemicals to kill fast-growing cells in your body
    a. Echo interrogation
        i) Decreased ventricular function
        ii) Pericardial effusion and constriction
        iii) Valvular disease
        iv) Heart failure

E. Rheumatic Heart Disease: Resulting From Inadequately Treated Strep Throat or Scarlet Fever

1. Rheumatic fever causes inflammation, especially of the heart, blood vessels and joints
2. Rheumatic fever is caused by Group A streptococcal bacterial infections
3. Cardiac manifestations
    a. Inflammation of the heart or heart valves
    b. Myocarditis
    c. Mitral stenosis (most common)
    d. Aortic stenosis

e. Pulmonary stenosis

f. Tricuspid stenosis (rare)

Section XIV: Pulmonary Arterial Hypertension (PAH)

1. Identify the types and causes of pulmonary arterial hypertension and pulmonary hypertension in pediatric patients
2. Describe the pathophysiology and progression of pulmonary arterial hypertension, including vascular changes and clinical implications
3. Recognize the echocardiographic methods used to evaluate pulmonary arterial hypertension and interpret relevant findings
4. List the clinical signs, symptoms, and complications associated with persistent fetal circulation and Eisenmenger syndrome
5. Summarize medical and surgical management strategies for pulmonary arterial hypertension and its associated conditions

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XIV: PULMONARY ARTERIAL HYPERTENSION (PAH)

A. Pulmonary Hypertension

1. Can be present with most forms of congenital heart disease
2. PAP elevates in response to increased blood flow volume of pressure as the pulmonary vascular resistance increases
3. Etiology
   a. A mean pulmonary artery pressure of > 25 mmHg at rest or > 30 mmHg with exercise in the absence of increased left-sided pressures
   b. Pulmonary endothelial dysfunction promotes a triad of vasoconstriction, cell proliferation and thrombus through the mediators such as thromboxane A2, endothelian 1 and Serotonin
   c. Under normal circumstances these are counterbalanced by prostacyclin, nitric oxide and vasoactive intestinal peptides
   d. The end result of this formulation is irreversible vascular changes with intimal fibrosis, pulmonary arteriolar occlusion and plexiform lesions
   e. The patient develops pulmonary hypertension (PHTN) and eventually pulmonary vascular obstructive disease
4. Types
   a. Primary or idiopathic
   b. Type I – congenital heart disease associated with a large ventricular septal defect and unobstructed pulmonary outflow tract or shunts at great vessel level
      i) Can also be seen with patent ductus arteriosus
      ii) Aortopulmonary window
      iii) Truncus arteriosus
   c. Type II – congenital pulmonary venous abnormalities encountered either in the newborn period with total anomalous pulmonary venous drainage, or in the form of congenital pulmonary vein stenosis
   d. Type III – absence of a mediastinal left or right pulmonary artery may be associated with pulmonary hypertension, particularly when the other lung is exposed to an intracardiac or extracardiac shunt
      i) PAH is often out of keeping with the magnitude of left-to-right shunt
   e. Type IV – PAH may be secondary to upper airway obstruction
      i) Down's Syndrome
      ii) Sleep apnea

iii) Obesity

iv) Interstitial fibrosis

v) Prior severe hyaline membrane disease

vi) Diaphragmatic hernia

vii) Chronic exposure to high altitudes

f. Type V – PAH in the newborn period in the absence of associated structural heart disease

g. Type VI – PAH secondary to systemic atrioventricular valve regurgitation or left ventricular systolic dysfunction

h. Type VII – PAH secondary to left-sided obstructive lesions

i) Coarctation of the aorta

ii) Cor triatriatum

iii) Mitral stenosis

iv) Hypoplasia of the systemic left ventricular is seen in a newborn period

i. Type VIII – pulmonary embolic disease encountered in infants with chronic venous lines

j. Type IX – postoperative period in newborns and infants with structural heart disease who have undergone repair of large intracardiac left-to-right shunt

i) Secondary

ii) Familial (genetic component)

4. Echocardiographic evaluation

a. Indirect measurement of pulmonary artery pressure

i) Interventricular septal shape

ii) Size and function of both ventricles

b. Normal anatomy may show a suprasystemic right ventricle; results in the interventricular septum bowing into the left ventricle

i) Size of right atria

ii) Right ventricular function

iii) Size of main pulmonary artery

c. Systolic time intervals

i) Pre-ejection period (PEP) – measured from the onset of the “q” wave to the beginning of the pulmonary artery acceleration

ii) Ejection time (ET) – related to stroke volume and inversely related to contractility

iii) Acceleration time (AT) – interval measured from the onset to the peak ejections

d. PV motion

i) Prominent “a” wave (from M-mode of pulmonary valve) is absent in PHTN

e. Tricuspid and pulmonary valve time intervals

i) Time interval between PV closure and tricuspid valve opening (isovolumic) has been used to predict indirectly systolic right ventricular pressure

f. Tricuspid regurgitation

i) Peak velocity

g. Right ventricular systolic pressure

i) Peak velocity of TR + right arterial pressure (RAP)

h. Pulmonary regurgitation for pulmonary artery end-diastolic pressure (EDP)

i) Indirect measurement of PA pressure

- Pulmonary artery EDP = 0.49 x PA systolic pressure (from peak TR gradient)

i. In the case of a patient with PHTN with a ventricular septal defect

i) Continuous wave Doppler across the ventricular septal defect with angle that is maintained parallel from the VSD jet to the Doppler beam (color flow may also be helpful with this technique)

ii) Pulmonary artery hypertension with a ventricular septal defect and a right ventricular outflow tract

j. Patent arterial duct and pulmonary artery pressure measurement

i) Continuous wave of patent ductus arteriosus PDAO flow, trace from R wave to R wave

k. Measurement of pulmonary artery capacitance by Doppler

i) Right ventricular stroke volume (mL) = PV diameter $(cm^2)^2$ x VTI

ii) Pulmonary compliance = RV stroke volume (mL)/4 x (TR2 – PR2)

l. The impact of pulmonary artery hypertension on the right ventricle

i) Myocardial performance index (Tei index) – measurement of global ventricular function

ii) Right ventricular fractional shortening-measurement of end diastolic volume – end systolic volume/end diastolic volume x 100

iii) Tissue Doppler

- Velocity of tricuspid valve inflow E' wave
- Velocity of right ventricle e-prime wave

iv) Velocity of right ventricular wave (Systolic contraction)

m. Strain, strain rate of right ventricle

B. Persistent Fetal Circulation (Persistent Pulmonary Hypertension)

1. Pulmonary hypertension that leads to right-to-left shunt of the patent ductus arteriosus and patent foramen ovale in the absence of congenital heart disease
2. Increased pulmonary artery pressures
3. Cyanosis is present
4. Presents with full-term babies who had a difficult birth in which a baby receives an inadequate amount of oxygen during delivery

a. Conditions such as infections or birth asphyxia

b. Rare condition usually seen in newborns with respiratory distress syndrome, overwhelming sepsis, other aspiration syndromes and intrauterine hypoxia and ischemia

5. Results in pulmonary vasoconstriction and increased pulmonary vascular hypertrophy and decreased area of the pulmonary vascular bed

a. Limited flow through the pulmonary vascular bed decreases the body's supply of oxygen

b. Life threatening

6. Etiology

a. Prevalence of one/1500 live births

b. More common in males

c. More frequent in higher altitudes

d. Most cases of persistent fetal circulation result in complete recovery of death

7. Long term effects

a. Chronic lung disease

b. Cerebral infarction resulting in specific motor and/or cognitive deficits

c. Hearing loss

d. May be an association of sudden infant death syndrome

8. Presents

a. Right after birth

b. When the right arm and head remain pink, lower body is cyanotic

c. Pan cyanosis

d. Tachypnea and tachycardia

e. Low O_2 sats

f. Congestive heart failure

g. Systemic hypotension

h. PHTN

i. Loud second heart sound (P2)

9. Echo findings

a. Right to left shunts for patent foramen ovale, ventricular septal defect and patent ductus arteriosus

b. Severely elevated right heart pressures

10. Treatment

a. Oxygen therapy and ventilators

b. Extracorporeal membrane oxygenation (ECMO)

c. Pulmonary vasodilators

d. Treat cognitive heart failure

e. Watch carefully for signs of improvement and/or deterioration

C. Eisenmenger Syndrome

1. A left-to-right shunt that becomes a bidirectional or right-to-left shunt due to increased pulmonary vascular resistance and severe elevation of PA pressure

a. Caused by a large shunting defect

2. Etiology

a. Presents in about 3-10% of those with congenital heart disease

b. Progressive right heart volume overload

3. Shunting defects that cause Eisenmenger's can be categorized into two groups:
   a. Pre tricuspid valve
      i) Arterial septal defect
      ii) Common atrium
      iii) Partial anomalous pulmonary venous return
      iv) Total anomalous pulmonary venous return
   b. Post tricuspid valve
      i) Aortopulmonary window
      ii) Complete atrioventricular septal defect
      iii) Patent ductus arteriosus
      iv) Ventricular septal defect
      v) Single ventricle
      vi) Conotruncal anomalies including truncus arteriosus, tetralogy of Fallot and double outlet right ventricle
4. Pathology and pathophysiology
   a. The right atrium, right ventricle and main pulmonary artery are dilated
   b. The left atrium and left ventricle are unchanged
   c. Intimal thickening and fibrosis of pulmonary vasculature
5. Signs and symptoms
   a. Chest pain (15-40%)
   b. Syncope (10-40%)
   c. Hemoptysis (10-33%)
   d. Progressive overtime
   e. Cyanosis
   f. Digital clubbing
   g. Progressive weakness
   h. Dyspnea
   i. Pulmonary hypertension
6. Further progression
   a. Ventricular failure
   b. Edema
   c. Elevated venous pressure
   d. Liver distension
   e. Hemoptysis
   f. Erythrocytosis, thrombocytopenia and elevated risk of thrombus
7. Other clinical presentations include:
   a. Hyperuricemia, pigment gallstones, orthopedic issues and brain abscess from the intracardiac shunt

8. Cardiac testing
    a. Electrocardiogram (EKG)
        i) Right ventricular hypertrophy
        ii) Biventricular hypertrophy
        iii) Atrial flutter
        iv) Chest x-ray
            - Right atrial enlargement and right ventricular enlargement
        iv) Echocardiogram
        v) Cardiac catheterization
9. Echocardiographic evaluation
    a. Absent “a” dip of pulmonary valve
    b. Mid-systolic notch (‘flying W’ across the pulmonary valve)
    c. Dilated right atrium/dilated right ventricle
    d. Systolic flattening of interventricular septum
    e. Decreased right ventricular ejection time
    f. Increased right ventricular pre-ejection period
    g. Evaluate the right and left ventricular thickness dimensions and systolic function
    h. Color flow, continuous wave Doppler, pulsed wave Doppler of affected shunts to determine the presence and severity of shunts (ie: ASD, VSD, PDA)
    i. Retrograde flow in ascending aorta
    j. Determine the presence and severity of pulmonic regurgitation
    k. Determine the presence and severity of Tricuspid regurgitation
    l. Saline contrast study
    m. Qp:Qs
    n. Determine the presence and severity of aortic coarctation
        i) Determine the presence of ventriculocoronary communications
        ii) Note the hypoplastic or morphologic LV, aortic valve hypoplasia, stenosis or atresia if present
        iii) Note the hypoplastic or morphologic LV, mitral valve hypoplasia, stenosis or atresia if present
10. Postoperative echocardiographic evaluation
    a. Determine the adequacy of the Norwood procedure
    b. Determine the adequacy of cardiac transplantation
11. Medical treatment
    a. Treat congestive heart failure medically
    b. Anticoagulation
12. Surgical treatment
    a. Heart-lung transplant

Section XV: Syndromes Associated with Congenital Heart Disease

1. Identify common genetic syndromes linked to congenital heart disease
2. Recognize key cardiac defects associated with specific syndromes
3. Understand the increased risk of chromosomal abnormalities such as Trisomy, 13, 18 and 21
4. Correlate clinical features of syndromes with their characteristic cardiac anomalies
5. List the most frequent congenital heart defects encountered in each syndrome

XV: SYNDROMES ASSOCIATED WITH CONGENITAL HEART DISEASE

A. Syndromes

1. CHARGE syndrome (50% risk)
    a. Tetralogy of Fallot
    b. Ventricular septal defect
    c. Atrioventricular septal defect
    d. Coarctation
    e. Atrial septal defect
    f. Tricuspid atresia
    g. Double outlet right ventricle
    h. Right aortic arch
2. DiGeorge syndrome (22g) (84% risk)
    a. Ventricular septal defect
    b. Coarctation
    c. Truncus arteriosus
    d. Tetralogy of Fallot
    e. Transposition of the great arteries
    f. Double outlet right ventricle
    g. Interrupted aortic arch
3. Ehlers Danlos syndrome
    a. Ascending aorta aneurysm
    b. Coarctation
    c. Interrupted aortic arch
    d. Atrial septal defect
    e. Ventricular septal defect
    f. Dextrocardia
    g. Tetralogy of Fallot
    h. Bicuspid aortic valve
    i. Bicuspid tricuspid valve
4. Loeys dietz syndrome
    a. Aortic aneurysms
    b. Dissection of the aorta
    c. Patent ductus arteriosus

d. Atrial septal defect
e. Bicuspid AV

5. Marfan syndrome (95% risk)
   a. Dilated aortic root and/or dilated aorta
6. Noonan syndrome (65% risk)
   a. Pulmonary stenosis
   b. Ventricular septal defect
   c. Atrial septal defect
   d. Interrupted aortic arch
   e. Coarctation
   f. Total anomalous pulmonary venous return
   g. Hypertrophic cardiomyopathy
   h. Aortic stenosis
   i. Tetralogy of Fallot
7. Scimitar syndrome
   a. Partial anomalous pulmonary venous return
   b. Absent right PA
   c. Atrial septal defect
   d. Ventricular septal defect
   e. Tetralogy of Fallot
8. Shone syndrome
   a. Supravalvular mitral membrane (SVMM)
   b. Parachute MV
   c. Subaortic stenosis (membranous or muscular)
   d. Coarctation of the aorta
9. Tuberous sclerosis
   a. Rhabdomyoma
   b. Angioma
   c. Coarctation
   d. Interrupted aortic arch
10. Turner syndrome
    a. Coarctation of the aorta
    b. Bicuspid aortic valves
    c. Atrial septal defect
11. VACTERL syndrome (50% risk)
    a. Hypoplastic left ventricle
    b. Ventricular septal defect
12. William syndrome (100% risk)

a. Supravalvular aortic stenosis
b. Peripheral pulmonic branch stenosis
c. Interrupted aortic arch
d. Tetralogy of Fallot
e. Coarctation

B. Chromosomal Abnormalities Associated with Congenital Heart Disease

1. Trisomy 13 (Patau syndrome) (90+% risk)
   a. Ventricular septal defect
   b. Atrial septal defect
   c. Dextroposition of the great arteries
   d. Hypoplastic left heart syndrome
   e. Tetralogy of Fallot
   f. Atrioventricular septal defect
   g. Coarctation of the aorta
   h. Interrupted aortic arch (IAA)
2. Trisomy 18 (Edwards syndrome) (99+% risk)
   a. Ventricular septal defect
   b. Atrial septal defect
   c. Pulmonary stenosis
   d. Double outlet right ventricle
   e. Atrioventricular septal defect
   f. Coarctation of the aorta
   g. Interrupted aortic arch
3. Trisomy 21 (Down's syndrome) (50% risk)
   a. Ventricular septal defect
   b. Atrial septal defect
   c. Tetralogy of Fallot
   d. Atrioventricular septal defect
   e. Coarctation of the aorta
   f. Interrupted aortic arch
   g. Pulmonary valve stenosis

Section XVI: Surgical Corrections of Congenital Heart Disease

1. Identify common surgical procedures for congenital heart defects and their primary indications
2. Describe the echocardiographic assessments required before and after each surgical intervention
3. Recognize key postoperative complications and their clinical management
4. Understand the rationale and expected outcomes of palliative and corrective cardiac surgeries
5. Summarize the goals and steps of specialized procedures

XVI: SURGICAL CORRECTIONS OF CONGENITAL HEART DISEASE

A. Rashkind Balloon Septostomy

1. Most commonly performed atrial septostomy
2. Performed to improve atrial saturation by mixing the blood
3. Performed in the first hours of day of life
4. Performed for the following: dextro-transposition of the great arteries (D-TGA), total anomalous pulmonary venous return, valvar atresia, hypoplastic left heart syndrome, restrictive PFO
5. Echo interrogation
    a. Apical right sternal border and subcostals
    b. Can be done under echo guidance in cath lab
    c. Examine atrial anatomy
    d. Assess size of defect before and after
    e. Assess direction and type of restriction
    f. Assess for pericardial effusion

B. PDA Repair

1. The PDA is surgically ligated or titanium clips are used for closure
    a. Echo assessment
        i) Parasternal long and short axis, apical, right sternal border and subcostal view
            - Preop
                - Assess PDA (size, length, direction, hemodynamic significance)
            - Postop
                - Assess residual shunting, LPA and DAO laminar flow
        ii) PDA device (ductal occludder, piccolo, etc.)
        iii) Parasternal long and short axis, apical, right sternal border and subcostal view
            - Preop
                - Assess PDA (size, length, direction, hemodynamic significance)
            - Postop
                - Assess residual shunting, LPA and DAO obstruction

C. ASD Repair

1. Can be delayed till patient is symptomatic or older
2. Risk is general anesthesia
3. Pericardium or dacron patch or primary closure

a. Amplatzer device

4. Echo assessment

a. Apical, right sternal border and subcostal views

i) Preop

- Assess ASD (number, location, size, direction), pulmonary venous anatomy and right heart

ii) Postop

- Assess residual shunting, pulmonary vein flow, MV impingement, SVC flow, right heart and pericardial effusion

D. VSD Repair

1. Patient symptoms of heart failure and failure to thrive
2. Risk: incomplete closure, interrupted conductive tissue, acquired valve regurgitation
3. Dacron patch
4. Echo findings

a. Parasternal long and short axis, apical, right sternal border and subcostals

i) Preop

- Assess ventricular septal defect (VSD) (number, location, size, direction), valve function

ii) Postop

- Assess residual shunting, aortic valve function and pericardial effusion

E. AVSD Repair

1. Repair for PHTN and heart failure
2. Risks: complete heart block, residual AV valve regurgitation, incomplete closure, subaortic stenosis due to accessory AV valve tissue
3. Patch closure of septal defects and suture repair of cleft MV
4. Occurs at 3 to 6 months
5. Echo assessment

a. Parasternal long and short axis, apical, right sternal border and subcostal view

i) Preop

- Assess AV canal defect, identify the type of common AV valve (Rastelli type), assess for regurgitation

ii) Postop

- Evaluate for residual shunting, cleft MV, regurgitation and LVOTO

F. Tetralogy of Fallot Repair

1. Risks: low, wide-open PI, residual VSD, residual stenosis, AI due to root dilation, AV block
2. Common to skip palliative repair unless PA branches are hypoplastic
3. Performed at 4-12 months
4. Repair: resection of RV muscle bundles, ASD/VSD patch repair, PDA closure and established PA flow (widening and patched or conduit placement)

5. Echo assessment
    a. Parasternal outflow and short axis, apical, right sternal border and subcostal view
        i) Preop
            - Identify severity of RVOTO, assess PV and type of flow, coronary arteries, branch arteries, ventricular septal defect (location, size, number, direction) and arch situs
        ii) Postop
            - Evaluate for residual stenosis, PI, VSD, branch arteries and RVSP

G. Truncus Arteriosus Repair
1. The section of the PA is removed from the trunk
2. The main trunk is repaired with sutures
3. A valved conduit is attached to the branch pulmonary arteries
4. VSD is patched

H. Coarctation of the Aorta Repair
1. Coarctation excision with end-to-end anastomosis
2. Patch enlargement of the narrowed segment
3. Subclavian flap repair with closure of the PDA
4. Balloon angioplasty with or without stent placement is recommended for adults

I. Total Anomalous Pulmonary Venous Repair
1. Create a wide anastomosis between the LA and pulmonary vein confluence and the pulmonary vein is sutured into the LA
2. Vertical veins are ligated
3. PDA is ligated
4. ASD is patch closed
5. Echo assessment
    a. Look for pulmonary vein stenosis

J. Cardiac Transplant
1. Performed for cardiomyopathy, single ventricle palliation failure, hypoplastic left heart syndrome palliation failure
2. Risks: rejection, coronary artery disease, sudden death
3. Placed on donor wait list. Must match blood type and weight
4. Connected to remaining portions of the recipient heart vena cava and atria
5. Biopsy is gold standard for rejection
6. Echo assessment
    a. PLAX, PSAX, apical, suprasternal notch (SSN) and subcostals
        i) Biweekly for first two months then monthly for the first year
        ii) Assess for rejection: effusion, valve regurgitation, ventricle systolic and diastolic failure, increased wall thickness and increase dimension

K. Blalock-Thomas-Taussig Shunt
1. Provides supplemental pulmonary blood flow

2. Performed for:
    a. Severe cases of tetralogy of Fallot
    b. Double outlet RV with severe PS
    c. Pulmonary atresia
    d. Hypoplastic left heart syndrome (as part of Norwood procedure)
    e. Single ventricle hearts with severe PS or pulmonary atresia

L. Modified BTTS
1. Synthetic or biologic interposition tube (usually 3mm to 5mm in diameter) placed between a subclavian artery and a PA
2. Echo assessment
    a. Interrogate in SSN
    b. Evaluate graft for narrowing thrombus
    c. Evaluate PA branches
    d. Diastolic flow reversal in the abdominal descending aorta

M. Waterston Shunt
1. Surgical aorto-pulmonary shunt by creating a fistula between the ascending aorta to the RPA to increase pulmonary blood flow in cyanotic heart disease
2. No longer used as a primary palliation
3. Performed for tetralogy of Fallot, tricuspid atresia, PA atresia
4. Echo assessment
    a. PSAX, SSN, PLAX, outflow, apical and subcostal view

N. Central Shunt
1. Palliative procedure in which the ascending aorta is attached to the PA via gortex graft
2. Indications are the following: pulmonary atresia, TOF, TA

O. Damus-Kaye-Stansel
1. Performed for double inlet left ventricle (DILV), single ventricle with sub aortic obstruction, tricuspid atresia with transposition, hypoplastic left heart syndrome (HLHS), double outlet right ventricle (DORV) with malposition, subpulmonic ventricular septal defect and coarctation of the aorta
2. Suturing together of PA to ascending aorta above the valve opening, ventricular septal defect patch closure and RV to PA conduit
3. Purpose is to provide systemic blood flow from both tracts
4. Echo findings
    a. At cardiac base, apical and subcostal view
        i) Postop
            - Evaluate anastomosis of aorta and pulmonary outflows for stenosis, for conduit obstruction, look for residual shunt

P. Palliative-Norwood Procedure
1. Surgical reconstruction of the aortic arch for cases of hypoplastic left heart syndrome or single ventricle with aortic hypoplasia or atresia
2. Aortic arch is augmented using biologic patch material, coarctation shelf and ductal tissue

are resected

3. Atrial septectomy
4. Modified Blalock-Taussig-Thomas shunt (BTTS) – systemic to pulmonary shunt is performed as secondary component of operation
5. Pulmonary trunk is transected from the branch pulmonary arteries
6. Native ascending aorta and pulmonary trunk are combined (DKS) and anastomosed to the reconstructed “neo-aortic” arch
7. Sano modified PA conduit may be used instead of a BT shunt
    a. Placed in the systemic ventricle to the PA

Q. Sano Modified Conduit

1. Surgical placement of a synthetic or biologic conduit between a ventricle and the PA bifurcation as a source of pulmonary blood flow
2. Used in first stage of single-ventricle palliations, hypoplastic left heart syndrome, hypoplastic right heart
3. Conduit diameter ranges between 3mm and 5mm
4. Echo assessment
    a. Apical and subcostal views
    b. Assess for narrowing and thrombus at anastomosis
    c. Assess main and branch pulmonary arteries

R. Hybrid

1. Goal of hybrid is to open blood flow through the PDA, balance blood flow through the lungs and body, unlock atrial septum
    a. Placement of bands around the right and left pulmonary arteries
    b. Stenting of the PDA
    c. Possible arterial septostomy if needed
    d. Echo assessment
        i) Parasternal, apical, subcostal and SSN views
        ii) Assess PDA stent for unobstructed flow
        iii) Assess branch PDA bands for restrictive flow
        iv) Assess atrial septum for unrestrictive flow

S. Bidirectional Glenn aka Hemi-Fontan

1. Anastomosis between the SVC and the RPA
2. RPA remains connected to PA and LPA
3. Directs flow to both lungs
    a. Can be performed without bypass
4. May develop aortopulmonary collaterals
5. Performed for hypoplastic left heart syndrome (2^{nd} stage), single ventricle (2^{nd} stage), pulmonary stenosis and atresia, tricuspid atresia
6. Echo assessment
    a. Supraclavicular/SSN short axis views

b. Look for bilateral SVC, systemic venous anomalies and normal branch PAs
c. Assess for narrowing and thrombus at anastomosis
d. Assess for SVC syndrome
e. Assess branch PAs
f. Look for low velocity phasic flow from shunt to PA

T. Fontan Repair

1. Performed for tricuspid atresia, HLHS, HRHS, single ventricle
2. Inferior cavo-pulmonary connection (often performed after a bidirectional Glenn)
3. Routing of IVC blood flow to the pulmonary arteries in cases of single ventricle physiology
4. Final stage for completion of single ventricle palliation
5. All systemic blood bypasses the right heart and is directed to the lungs
6. Types
   a. “Classic” atrio-pulmonary connection (no longer performed)
   b. Direct connection of RA to pulmonary arteries with PFO closure
   c. Lateral tunnel: intra-atrial baffle
   d. Extra-cardiac conduit uses conduit
7. Echo assessment
   a. Supraclavicular/SSN short axis, high right parasternal
   b. Assess for narrowing and thrombus at anastomosis
   c. Assess main and branch PAs
   d. Look for low velocity phasic flow from shunt to PA

U. Pulmonary Artery Band

1. Palliative procedure used to decrease pulmonary blood flow
2. Performed via left thoracotomy
3. Performed for AV septal defect, VSD, single ventricle
4. Possible complications
   a. Band migration causing flow obstruction to one or both proximal branch PAs
   b. Band not tight enough
   c. Increasing valve regurgitation
   d. Increasing RVH
   e. PA aneurysm
   f. Residual PA stenosis
   g. Valve leaflets destruction
5. Echo assessment
   a. Suprasternal notch, parasternal outflow, apical and subcostal views
   b. Bright white reflective band in the PA not associated with leaflets
   c. Band is removed when corrective repair achieved
   d. High systolic velocity similar to PS

V. Rastelli Procedure

1. Repair of double outlet right ventricle, dextro-transposed great arteries with ventricular septal defect and truncus arteriosus
2. Risks: PI and conduit stenosis
3. LV outflow is routed through the ventricular septal defect into the RV to the remote aorta via baffle patch
4. Placement of RV to PA hemograft or conduit
5. Procedure may include LeCompte maneuver of the pulmonary arteries
6. Echo assessment
    a. Apical and subcostal views
    b. Preop
        i) Assess the relationship of aorta to VSD, heart disease
    c. Postop
        i) Evaluate conduit stenosis/insufficiency, thrombus or pseudoaneurysm, residual ventricular septal defect shunting

W. Mustard and Senning Procedure

1. Older repair of dextro-transposition of the great arteries
2. Atrial switch operation
    a. Maybe used for congenital transposition of the great arteries
3. Rarely performed
4. Mustard replaced Senning
    a. Senning is a tissue from the RA used to create the baffle
    b. Mustard uses pericardium or prosthetic material
5. Risks: baffle obstruction, atrial arrhythmias, RVOTO or LVOTO, RV failure, sudden death
6. Large patch creates and interatrial baffle
7. Redirects systemic and pulmonary venous inflows at the atrial level. Systemic venous return is directed across the atrial septum to the LV to PA. Pulmonary venous return is rerouted to the right ventricle to aorta
8. Echo assessment
    a. PSAX, apical and subcostal views
        i) Postop
            - Evaluate pulmonary venous return, systemic venous baffles for leaks or obstructions, LVOT for obstruction, ventricular function

X. Jatene Operation (Arterial Switch)

1. Indicated for dextro-transposition of the great arteries
2. Risks: supravalvular pulmonary or aortic stenosis, coronary artery stenosis
3. Aorta and pulmonary arteries are transected
4. Ascending aorta is anastomosed to native pulmonary root
5. Pulmonary trunk is anastomosed to native aortic root
6. Coronary arteries are re-implanted to the root of the “neo-aorta”

7. Echo assessment
    a. Parasternal long, outflow, short axis, apical, right sternal border and subcostal view
        i) Preop
            - Measure PA and aorta, assess semilunar valve anatomy and coronary artery anatomy
        ii) Postop
            - Evaluate ventricular outflows, supravalvular stenosis, coronary arteries and residual atrial shunt

Y. LeCompte Maneuver

1. A surgical method of positioning the branch pulmonary arteries anterior to the aorta
2. Used during Jatene ASO when great artery relationship is anterior/posterior
3. Pulmonary artery branches “straddle” the ascending aorta
4. Used in rare cases of PA compression of airways (TOF with absent pulmonary valve syndrome)
5. Echo assessment
    a. Evaluate for pulmonary artery branch stenosis

Z. Konno Procedure

1. Performed for subaortic stenosis and hypoplastic aortic valve
2. Risks: heart block
3. Aortic valve is removed and small incision is made in ventricular septum (ventriculoplasty)
4. The incision is widened with a patch to relieve obstruction
5. Aortic valve is replaced with a mechanical prosthesis (Konno-Rastan), aortic hemograft, or patient’s own pulmonic valve (Ross-Konno)
6. In some cases a pacemaker is placed
7. Echo assessment
    a. PLAX, apical and subcostals
        i) Postop
            - Evaluate for ventricular level shunting, aortic valve and residual left ventricular obstruction

AA. Ross Procedure

1. Used for severe congenital aortic valve stenosis and/or regurgitation
2. Surgical replacement of aortic root and valve with native pulmonary trunk (pulmonary autograft)
3. Placement of RV to PA homograft or conduit
4. May be used in conjunction with the Konno procedure (i.e., Ross-Konno)
5. Echo assessment
    a. PLAX, apical and subcostal views
        i) Preop
            - Assess semilunar annular dimensions
        ii) Postop

- Evaluate RV conduit for stenosis, neo-aorta for stenosis/insufficiency, long term aortic root dilation is common

BB. Warden Procedure

1. Surgical repair of sinus venosus with partial anomalous venous return to the SVC
2. The SVC is transected above the level of the anomalous pulmonary venous connection
3. The SVC "stump" on the atrial side is sutured closed
4. An intra-atrial baffle is placed from the SVC orifice to the ASD, effectively routing the anomalous pulmonary venous flow to the LA
5. The SVC is then anastomosed anteriorly to the RAA

CC. CONE Procedure for Ebstein Anomaly

1. Extensive reconstruction of the tricuspid valve including:
   a. Detaching tethered septal leaflet
   b. Creating an effective TV annular region
   c. Re-suspending TV apparatus
   d. Pilication of excessive atrialized RV

DD. Postop Complications

1. Post pericardiotomy syndrome
   a. Autoimmune response from opening the pericardial sac
   b. Pericardial effusion and tamponade may result
   c. Clinical signs
      i) Fever, chest pain, SOB, pulsus paradoxous
   d. Echo evaluation
      i) Degree of pericardial effusion, signs of tamponade, assess for pleural effusion
   e. Treatment
      i) Steroid treatment
      ii) Anti-inflammatory medications
      iii) Pericardiocentesis
2. Postperfusion syndrome
   a. Patients experience neuro deficits following bypass
   b. Presents 3 to 6 weeks after surgery
   c. Symptoms
      i) Attention disorder, loss in concentration, short term memory loss, fine motor function and speed of mental and motor responses
   d. Theorized that it's caused by tiny debris and air bubbles that enter the brain via cardiopulmonary bypass
3. Paralyzed diaphragm
   a. Often a cause of dyspnea mainly on exertion
   b. Diaphragm contracts on inspiration normally
   c. Paralyzed may cause right atrial compression

i) Absence of pericardial effusion or mass

ii) Symptoms are pronounced with exertion

iii) In Glenn and Fontan procedures, the loss of diaphragm function is also associated with redistribution of pulmonary flow away from the affected side

Section XVII: Fetal Echocardiography

1. Identify optimal timing and indications for fetal echocardiography
2. Describe the key fetal heart views and assessment techniques used in routine and detailed scans
3. Recognize maternal and fetal risk factors for congenital heart disease
4. List common teratogens and associated cardiac defects
5. Explain the normal fetal cardiovascular physiology and circulation
6. Summarize the goals and steps of the fetal echocardiogram examination
7. Interpret biometry measurements and Doppler findings relevant to cardiac assessment
8. Discuss the clinical impact and potential prenatal interventions enabled by fetal echocardiography

XVII: FETAL ECHOCARDIOGRAPHY

A. Timing of Fetal Echo

1. Can be performed as early as first trimester
2. The best time to perform a fetal echo is between 18 – 22 weeks
3. After 22 weeks, cardiac structures are larger and more visible
4. 3rd trimester is suboptimal due to decreased amniotic fluid, fetal bone ossification and suboptimal fetal position

B. Indications for Fetal Echo

1. Low risk patients will have a routine anatomy scan with incorporated heart views in second trimester
2. Views during an anatomy scan
   a. Abdominal visceral situs and cardia situs
   b. 4 chamber
   c. Aortic valve/left ventricular outflow track
   d. PV and right ventricular outflow track
   e. 3 vessel view
   f. Bicaval view
3. High risk patients will have a detailed fetal echocardiogram
4. Views during fetal echocardiogram
   a. Abdominal visceral situs and cardiac situs
   b. 4 chamber view
   c. Aortic valve/left ventricular outflow track
   d. PV and right ventricular outflow track
   e. 3 vessel view
   f. 3 vessel tracheal view
   g. Cardiac connections
   h. Cardiac function
5. Fetal echocardiograms performed in 1st trimester indications include:
   a. A previous child with severe cardiac disease
   b. Increased nuchal translucency
      i) Higher risk of chromosomal abnormalities
   c. Ductus venosus flow patterns “a” reversal

d. Tricuspid regurgitation

6. Maternal indication
   a. Family history of congenital heart disease
   b. Metabolic disorders (e.g. diabetes, phenylketonuria)
   c. Exposure to teratogens
   d. Exposure to prostaglandin synthetase inhibitors (e.g., ibuprofen, salicylic acid)
   e. Rubella infection
   f. Autoimmune disease (e.g. Sjögren's syndrome, systemic lupus erythematosus)
   g. Familial inherited disorders (ells van Creveld, Marfan's syndrome, Noonan's syndrome)
   h. In vitro fertilization
7. Fetal indications
   a. Abnormal obstetrical ultrasound screen
   b. Extracardiac abnormality
   c. Chromosomal or genetic abnormality
   d. Irregular heart rhythm
   e. Hydrops
   f. Increased first-trimester nuchal translucency
   g. Multiple gestation and suspicion of twins – twin transfusion syndrome
   h. Family history of congenital heart disease
8. Maternal conditions associated with congenital heart defects
   a. Maternal rubella (50%)
      i) Branch pulmonary stenosis
      ii) PDA
   b. Maternal diabetes
      i) Dextro-transposition of the great arteries (D-TGA), truncus, single ventricle
   c. Maternal phenylketonuria (PKU ~20%)
      i) Tetralogy of Fallot
      ii) Coarctation of the aorta
   d. Systemic lupus (20 – 40%)
      i) Complete heart block
   e. Maternal infection
      i) Dilated or hypertrophic cardiomyopathies
9. Maternal exposure to cardiac teratogens
   a. Increases the risk of congenital heart defect
   b. The specific risk of occurrence varies with length of exposure, types of exposure and the specific substance involved
   c. Drugs are commonly thought of a teratogens; however, other disease processes or external influences can also have teratogenic effects of the fetus, such as diabetes or

obesity

d. An increase in awareness of teratogens has decreased the risk to women in the reproductive age group

10. Ace inhibitors (5 – 10%)
    a. VSD
    b. ASD
11. Alcohol (50%)
    a. ASD
    b. VSD
12. Amantadine
    a. SV
    b. PA
13. Amphetamine (5 – 10% risk)
    a. ASD
    b. VSD
    c. TGA
14. Azathioprine
    a. PS
15. Barbituates
    a. IAA
    b. Coarctation
16. Carbamazepine
    a. ASD
17. Chlordiazepoxide
    a. Codine
18. Cortisone
    a. VSD
    b. Coarctation
19. Cyclophosphamide
    a. TOF
20. Cytarabine
    a. TOF
21. Daunorubicin
    a. TOF
22. Dextroamphetamine
    a. ASD
23. Diazepam
24. Dilantin (hydantoin) (10%)

a. ASD
b. VSD
c. PS
d. AS
e. TOF

25. Indomethacin (50%)
    a. Ductal constriction
    b. PFO restriction
26. Lithium
    a. Ebstein's anomaly
    b. Tricuspid atresia
    c. ASD
    d. Mitral atresia
    e. Dextrocardia
27. Oral contraceptives
28. Paramethadione
    a. TOF
29. Penicillamine
    a. VSD
30. Primidone
    a. VSD
    b. IAA
    c. Coarctation
31. Progesterone
    a. VSD
    b. TOF
    c. TA
32. Quinine
33. Retinoic acid (accutane) (50%)
    a. TGA
    b. TOF
    c. VSD
    d. IAA
34. Selective serotonin reuptake inhibitors (74%)
    a. PHTN
35. Thalidomide
    a. TOF
    b. TA

c. DORV
d. ASD
e. PA

36. Trifluoperazine
37. Trimethadione (~50%)
    a. VSD
    b. ASD
    c. PDA
    d. TOF
    e. HLHS
    f. DORV
    g. ASD
    h. PA
    i. TA
    j. AS
    k. PS
38. Vitamin D
    a. Supravalvular AS
39. Warfarin (~10%)
    a. PDA
    b. PS

C. Normal Fetal Cardiovascular System
1. Heart of 120 – 160 bpm
2. Lower arterial blood pressure
3. Lower peripheral resistance
4. Lower myocardial compliance and contractility
5. Fluid-filled lungs
6. The lungs do not provide gas exchange
7. The pulmonary vessels are vasoconstricted
8. Closer to term, the ventricle becomes more compliant

D. Goals of the Fetal Echocardiogram
1. Determine fetal position
2. Determine visceral/abdominal situs
3. Determine morphology and connections of atria and ventricles
4. Determine morphology, patency and flow of the great vessels
5. Examine the size and function of AV and semilunar valves
6. Determine heart rate and rhythm assessment
7. Obtain cardiac biometry and function assessment

E. Vascular Structures Most Important in Fetal Circulation

1. Ductus venosus – a fetal vessel connecting the umbilical vein to the inferior vena cava
2. Foramen ovale – communication between the right and left atria
3. Ductus arteriosus – communication between the descending aorta and the LPA

F. The Fetal Echocardiogram

1. The heart is assessed by transversed, sagittal and oblique views of the fetal heart
2. A segmental approach is used
   a. Begin with the fetus position in the womb
   b. Explain the location of the fetal head
      i) Breech (head is up)
         - Head located in the fundus
      ii) Cephalic/vertex with head down
         - Head is located closest to the cervix
      iii) Spine up
      iv) Spine down
      v) Facing left
      vi) Facing right
   c. Determine abdominal situs
      i) Situs solitus
         - Stomach on the left
         - Liver on the right
         - Abdominal aorta is to the left and anterior of the spine
         - Inferior vena cava is anterior and to the right of the abdominal aorta
      ii) Situs inversus
         - Stomach on the right
         - Liver on the left
         - Abdominal aorta is to the right and anterior of the spine
         - IVC is anterior and to the left of the abdominal aorta
      iii) Situs ambiguous
         - Visceral organs have no laterality (i.e. midline liver, stomach)
         - Juxtaposed great arteries
   d. Determine cardiac position and cardiothoracic position
      i) The normal position of the heart = interventricular septum (IVS) is at a 45-degree angle +/- 15 degrees from midline
      ii) Apex of the heart pointed
         - Levocardia
         - Mesocardiac
         - Dextrocardia

iii) Levoposition, detroposition

e. Transverse/oblique images

i) Fetal 4 chamber view

- Apical 4 chamber is obtained with the fetal chest in transverse
- Apex is towards or away from the transducer
- The septum should be parallel to the transducer

ii) Subcostal 4 chamber

- Fetal chest in transverse with the septum running horizontal

iii) 4 chamber assessment (2D, color, PW Doppler)

- AV leaflets opening
- AV annulus size
- AV offset = tricuspid valve slightly inferior
- Pulmonary venous connections (lower color scale)
- Atria or equal size
- Appendage if visualized
- Foramen ovale flap in the left atrium
- Ventricles are equal size
- Moderator band in RV
- Color Doppler to assess
    - Foramen ovale flow
    - Intact ventricular septum
    - Color will depend on apex position
        - ~ Blue or Red
    - Valvular or arch turbulence
    - Atrioventricular valve regurgitation
- PW Doppler to assess
    - Atrioventricular PW is performed at the leaflet tips in the ventricle
    - This doppler assesses the relaxation of the ventricle
    - The pattern consists of an E wave and an A wave
        - ~ E wave is the rush of blood into the ventricle as the inflow valve opens
        - ~ A wave corresponds to the atrial kick
        - ~ The ventricles are stiff in utero
        - ~ Reversed with low E wave and high A wave
        - ~ Peak velocities of this valve increase with gestation
        - ~ To look for the direction of the blood flow and velocities

iv) Sweep of the outflow tract

- Assess the valve opening

- Size of the aorta
- Criss crossing of the great vessels
- PA rises anteriorly from the RV
- Aorta rises posteriorly from the LV
- Assessment of intact septum

v) Left ventricular outflow tract

- From the 4-chamber angle towards fetus right shoulder
- Slight clockward turn
- Cardiac output = $0.785 \times (D)^2 \times$ LVOT VTI x HR
  - Normal = 60 – 120 ml for LV
  - Calculate both right and left
  - RV CO>LV CO
- High output – fetal anemia, AV malformations
- Low output – cardiomyopathy, heart block
- Spectral Doppler to assess aortic valve stenosis or insufficiency

vi) Right ventricular outflow tract

- From the 4-chamber angle towards fetus right shoulder
- Further clockward turn then LVOT
- RVOT assessment
  - Two branches
  - Valve patency
  - Size of the PA
  - Size of the pulmonary branches
  - Criss cross with aorta

vii) Three vessel view/3 vessel tracheal view

- From the 4-chamber angle/slide the transducer cephalad
- Assessment
  - Size difference between the PA, aorta, SVC
  - # of vessels
  - Direction of blood flow

viii) Short axis of the great vessel level

- Turn transducer in clockwise manner for a sagittal view of the thorax; this will result in short axis of the ventricles
- Angle slightly towards the fetus left shoulder for great vessels

ix) Short axis of the ventricles

- Turn transducer in clockwise manner for a sagittal view of the thorax; this will result in short axis of the ventricles

f. Sagittal views of superior and inferior vena cava aortic arch and ductal arch

i) Bicaval view

- Turn transducer in clockwise manner for a parasagittal view of the thorax; sweep to the right of the fetus
- Look for the patency and phasic flow pattern in the inferior vena cava & superior vena cava

ii) Aortic arch

- Align a sagittal view of the thorax and sweep the beam from the left shoulder to the right hemithorax of the fetus
- Look for the patency, size and flow pattern

iii) Ductal arch

- Align a sagittal view of the thorax and sweep the beam midline of the fetus
- Look for the patency, size and flow pattern

iv) Ductus venous

- Stay on a parasagittal view of the thorax; look for umbilical insertion
- Look for the patency, size and flow pattern
- Follow the inferior vena cava for ductus venous connection
  - Patterns are similar to caval flows

g. Doppler assessment

i) Three types of Doppler

- Cardiac Doppler
  - Inflows
  - Outflows
  - Doppler Tissue imaging
- Arterial Doppler
  - Ductus arteriosus
  - Pulmonary arteries
  - Aorta
  - Cerebral
  - Umbilical artery
- Venous Doppler
  - Umbilical vein
  - Ductus venosus
  - IVC/SVC
  - Hepatic vein

h. Fetal biometry measurements

i) Biometry measurements compared to expected value for the fetal age

ii) Falls into lower than 5^{th} percentile or greater than 95^{th} percentile

iii) Cardiac defect could cause a decrease in size

iv) Cardiac defect causes an increase in size

i. Biometry measurements include:
   i) Measurement of femur length
   ii) Head circumference and diameter
   iii) Cardiac to thoracic ratio
   iv) Abdominal circumference

j. Umbilical cord flow
   i) Number of vessels
   ii) Arterial flow
   iii) Venous flow should be monophasic

k. Methods of assessing rhythm
   i) M-mode of the fetal heart
   ii) PW Doppler of mitral inflow and aortic outflow

G. Cardiac Anomalies
   1. Cyanotic congenital heart disease
   2. Non-cyanotic congenital heart disease

H. Cardiac Arrhythmias
   1. M-mode through right atria and left ventricle
   2. Pulse Doppler of inflow and outflow valves

I. 3D Fetal Echocardiography

J. Pitfalls of Scanning in Fetal Echocardiography
   1. No detection of very small VSDs
   2. Coarctation of the aorta
   3. Total anomalous pulmonary venous return
   4. Pulmonic and aortic stenosis
   5. Pseudo pericardial fluid
   6. Pseudo thickening of the tricuspid valve
   7. Pseudo overriding of the aorta

K. Impact of Fetal Echocardiography
   1. Prenatal diagnosis allows for planned deliveries with access to obstetricians, neonatologists and pediatric intensivists
   2. Fetal echo can detect ductal dependent lesions

L. Prenatal Interventions in the Fetus with Cardiac Disease
   1. In-utero intervention
   2. Valvuloplasty for aortic stenosis/hypoplastic left heart for improvement of ventricular growth and valvular stenosis
   3. Valvuloplasty for pulmonary stenosis and pulmonary atresia with intact ventricular septum

Section XVIII: Stress Echocardiography

1. Identify the clinical indications and contraindications for stress echocardiography
2. Distinguish between exercise and pharmacologic stress echocardiography, including preparation, equipment and endpoints
3. Describe the hemodynamic and ischemic responses observed during stress testing, including wall motion analysis
4. Recognize the applications of stress echocardiography in diagnosing coronary artery disease, valvular abnormalities and cardiomyopathies
5. List the advantages and limitations of stress echocardiography as a diagnostic tool

XVIII: STRESS ECHOCARDIOGRAPHY

- A. Provides Hemodynamic Information on a Person's Active State
- B. Applied for Two Diagnostic Issues
  1. Impairment of myocardial perfusion
  2. Hemodynamic patterns during stress of cardiac pathology
- C. Indications
  1. Familial hypercholesterolemia
  2. Diabetes mellitus
  3. Chronic kidney disease
  4. Kawasaki disease
  5. Rheumatologic diseases
  6. Post cardiac transplant
  7. Hypertrophic cardiomyopathy
  8. Anomalous aortic origin of coronary arteries
  9. Arterial switch operation for TGA
  10. Cardiotoxicity
  11. Post Fontan operation
  12. Ross procedures
  13. PHTN
  14. Coronary ostia stenosis
  15. ALCAPA
  16. Obstruction from left heart structures
      - a. Valvular stenosis
      - b. Coarctation of the aorta
  17. Tetralogy of Fallot
  18. Anomalous coronary artery origins
  19. Clinical applications for stress echocardiography
      - a. Diagnose coronary artery disease
      - b. Determine extent of coronary artery disease
      - c. Evaluate LV function
      - d. Screen preoperative and postoperative

e. Assess valvular heart disease
f. Evaluate myocardial viability
g. Evaluation of hypertrophic obstructive cardiomyopathy (HOCM)/systolic anterior motion severity

D. Coronary Artery Disease
1. Ischemia
a. Oxygen supply demand mismatch
2. Sequence of ischemic response
a. Decrease perfusion
b. Decrease LV compliance
c. Increase LVEDP
d. Decrease LV contractility
e. ECG changes
f. Symptoms
3. Indications
a. CAD screening
b. Assess perfusion pre and post revascularization
c. Determine prognosis
d. Evaluate valvular heart disease
4. Contraindications
a. Aortic stenosis
b. Obstructive hypertrophic cardiomyopathy

E. Types of Stress Echocardiography
1. Exercise stress echocardiography
a. Patient preparation and electrocardiogram (EKG) lead placement
b. Indication
c. Relative contraindication
d. Stress testing system
i) Treadmill
ii) Bicycle
iii) Other
e. 12-lead electrocardiogram (EKG) system
f. Blood pressure device
g. Emergency crash cart
h. End point
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 electrocardiogram (EKG) system

f. Blood pressure device

g. Emergency crash cart

h. Pharmacological stress agents

i) Effects on heart

ii) Atropine to achieve target heart rate

i. End points

i) Clinical ischemia

ii) Blood pressure (hyper/hypotension)

iii) Sustained tachycardia

iv) Wall motion abnormality involving two or more segments

j. Predicted heart rate

k. Pacing

l. Corresponding perfusion zones of the major coronary arteries

i) LAD

ii) LCX

iii) RCA

m. Procedure

i) Baseline images

- PLAX
- PSAX
- Apical 4 chamber
- Apical 2 chamber
- Apical long axis

ii) Stress initiated

- Target heart rate

iii) Post exercise echocardiographic images

- Time limitations
- Patient position
- Time increments

- Digital image acquisition
  - Tissue harmonic imaging
- Contrast for LV opacification

n. Ischemic response

i) Wall motion analysis

- Absence of hyperkinesis
- Normal function
- Hypokinetic
- Akinetic
- Dyskenetic

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 thickness

G. Pitfalls

1. May precipitate atrial and ventricular arrythmias
2. Obesity makes difficult imaging
   a. Due to obesity, limitation may be with contrast may be contraindicated due to patient diagnosis (i.e. shunting)

Section XIX: Transesophageal Echocardiography

1. Describe the purpose and protocol of transesophageal echocardiography
2. Identify the equipment necessary and list common clinical indications to perform an exam

XIX: TRANSESOPHAGEAL ECHOCARDIOGRAPHY

A. Transesophageal Echocardiography (TEE)

1. Purpose
   a. Unobstructed window
   b. Higher resolution compared to TEE
2. Indications
   a. Used in the assessment of anatomy and function during congenital heart disease heart surgery (intraoperative) (both pre and postoperative)
   b. Assist in catheter placement
   c. Further assessment of congenital heart disease
   d. Assessment of hemodynamics and myocardial function
   e. Real-time monitoring of volume status and myocardial function
   f. Used in cath lab for assistance for placement of septal devices and safety in placement of interventional devices
   g. Review of prosthetic valves
   h. Review of intra-cardiac thrombus
   i. Assistance in the diagnosis and follow up for infective endocarditis
3. Contraindications
   a. The benefits of the procedures should outweigh the risks
   b. Relative contraindications primarily relate to the potential of esophageal trauma or airway compromise
   c. Risk with Down's syndrome with large tongue
   d. Care and risk after gastrostomy tube/button placement (with or without associated Nissen fundoplication) with avoidance of stomach transgastric or deep transgastric imaging
   e. Bleeding
4. Procedure
   a. Description
   b. Equipment
   c. Transducer types
      i) Monoplane
      ii) Biplane
      iii) Omniplane
      iv) Dimensional
   d. Transducer selection
      i) Weight and probe size
      ii) Micro multiplane probes for patients greater than 2.5kg

iii) Mini-Multiplane probe for patients greater than 3-3.5kg
- Adult probe (TEE) available for use in patients greater than 25kg
- 3D TEE adult probe available for use in patients greater than 30kg

iv) Each probe may be used safely below weight recommendations

e. Types of sedations

f. Patient preparation

i) Probe insertion should be performed with adequate sedation and anesthesia

ii) Recommendations for appropriate period of fasting

iii) Patient sedation
- Careful monitoring for complications is essential

g. Laboratory set up/clean up

h. Transducer care

i) Cleaning

ii) Rinsing and high-level disinfection should be performed after each use

iii) Storage

5. Advantages/limitations

6. Complications

7. Training and certification as outlined in the task force addressing pediatric cardiology fellowship training in non-invasive cardiac imaging

a. Supervised performance and interpretation of at least 50 TEE examinations in pediatric and adult congenital heart disease patients prior to independent TEE imaging

8. Physician maintenance of TEE skills

a. 25-50 annual TEE exams in pediatric and/or adult congenital heart disease (ACHD) patients

9. Physician certification

a. There is currently ne certification pathway specifically designed for the physician performing TEE for the pediatric or adult congenital heart disease patient

10. Imaging views

a. Mid esophageal (ME) 4 chamber-depth to include LV

i) 0 degree 4-chamber – retroflex
- Left atrium/right atrium
- Interatrial septum
- Left ventricle/right ventricle/interventricular septum
- MV (A3A2-P2P1) tricuspid valve, coronary sinus

ii) 0-10 degree anterflex to see aortic valve and left ventricular outflow tract (ME5 chamber)

iii) 50-70 degree ME mitral

iv) 80-100 degree 2-chamber

- LV size and function
- LA and LV appendage
- MV

v) 120-140 degree ME long axis

- LV size and function
- LA size
- Mitral and aortic valve
- Withdraw probe for ascending aorta

b. Mid-esophageal (ME) – decrease depth to optimize valves

i) 120-140 degree ME aortic valve LAX

ii) 90-110 degree ME ascending aortic valve LAX

iii) 90-110 degree ME ascending aortic valve SAX

iv) 0 degree ME right pulmonary veins

v) 90-110 degree ME left pulmonary veins

vi) 90-110 degree ME LAA (LAA/LUPV)

vii) 25-45 degree ME aortic valve SAX

viii) 25-45 degree ME RV in-out

ix) 50-70 degree ME mod bicaval TV

- Left atrium/right atrium
- Mid interatrial septum
- Tricuspid valve
- Superior vena cava/inferior vena cava
- Coronary sinus

iv) 90-110 degree ME bicaval

- Left atrium (LA)/right atrium (RA)
- Mid interatrial septum (IAS)
- Superior vena cava/inferior vena cava
- Coronary sinus

c. Transgastric (TE) views

i) 80-100 degree TG inferior vena cava/hepatic veins

ii) 0-20 degree TGs basal SAX left ventricle/right ventricle/inferior vena cava

- LV wall motion, thickness and dimensions
- RV size and function

iii) 0-20 degree TG mid SAX LV

- LV wall motion, thickness and dimensions
- RV size and function

iv) 0-20 degree TG mid apical SAX

- LV wall motion, thickness and dimensions
- RV size and function

v) 0-20 degree TG apical SAX

- LV wall motion, thickness and dimensions
- RV size and function
- Interventricular septum; apical muscular

vi) 120-140 degree TG long axis

- Mitral valve anatomy and function
- Color Doppler for MR
- Left atrial (LA) imaging and flow

vii) 120-140 degree transgastric LAX

- Anatomy and function
- Antegrade aortic flow with color Doppler

d. Deep transgastric (DTG) views

i) 0-20 degree DTG 5 chamber

ii) 50 degree DTG RVOT

iii) 80 degree DTG atrial septum (bicaval)

e. Upper esophageal views

i) 0-10 degree UE long axis

- Aortic arch and innominate vein

ii) 70-90 degree UE aortic arch SAX

- Aortic arch
- Innominate vein
- MPA, Branch PA's, PV

iii) 0-20 degree UE upper arch PA

- PA, branch PA's
- Mid ascending aorta

iv) 0-10 degree UE descending aorta SAX

v) 90-100 degree UE descending aorta LAX

11. 28 views for a complete pediatric/adult congenital TEE
    a. Modify each view according to anatomy and pathology
12. Probe manipulation
    a. Advance/withdraw
    b. Rotation (0 – 180 degree)
    c. Turn (right left/medial/lateral)
    d. Anteflex/retroflex
    e. Flex right/left

13. 3D TEE
    a. Provides further evaluation of valve size
    b. Provides further evaluation of pathology
    c. Provides further evaluation of mechanism and valve dysfunction
    d. Volumetric ejection fraction
    e. Quantitate assessment of RV function
14. Safety practices
    a. TEE demonstrates a favorable safety profile and the complication rate is low in children/adults with congenital heart disease
    b. Most complications are related to respiratory compromise or vascular compression
    c. Rare serious complications include esophageal perforation, gastric laceration and subglottic stenosis
    d. Recommendations for general anesthesia for children
    e. Recommendations for conscious sedation for the adult congenital heart disease patient
    f. Recommendations for general anesthesia for the following patients
        i) Fontan repair, unrepaired or palliated cyanotic cardiac patients or severe PHTN patients
15. Roles of the sonographer
    a. Assist in preparation of patient (monitoring, comfort)
    b. Optimization of images

Section XX: 3D/4D Echocardiography

1. Describe the structure and function of 3D/4D echocardiography transducers
2. Identify the advantages and limitations of 3D/4D versus 2D imaging modalities
3. Explain imaging methods and display techniques used in 3D/4D echocardiography
4. Recognize key clinical indications for 3D/4D echocardiography in congenital heart disease
5. Interpret anatomical landmarks and imaging views for common cardiac defects using 3D/4D echocardiography

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XX: 3D/4D ECHOCARDIOGRAPHY

A. 3D/4D Echocardiography

1. Transducer
    a. 3D matrix-array transducers composition
    b. Frequencies range are similar to 2D transthoracic and transesophageal transducers
    c. Piezoelectric elements are arranged in rows and columns to form a rectangular grid (matrix configuration) within the transducer
    d. Scan line propagates radially (y or axial direction), laterally (x or azimuthal direction) and in the elevation plane (z direction) to acquire a volumetric pyramid of data
2. Advantages
    a. Volumetric/functional 3D imaging
    b. Used for 3D analysis of ventricular or atrial volume and function
    c. Anatomic 3D imaging evaluation improves image quality
    d. Reduces artifacts
3. Imaging methods and display
4. Image acquisition
    a. Atrioventricular or semilunar valves
    b. Atrial septal defects
    c. Ventricular septal defects
    d. Outflow tracts
    e. Foreign bodies and masses
5. Multiplane imaging (multiplanar reconstruction MPR)
    a. Uses 2D to recognize space and time and drive 3D images
    b. Assessment of multiple views from the same cardiac cycle
    c. Useful for atrial fibrillation or other irregular arrhythmia, stress echo, evaluation of interventricular desynchrony
    d. Assessment of ASD size and rim length
    e. Size and shape of VSDs
    f. Inflow valve morphology and regurgitation
    g. Outflow tracts and arterial valves
6. Multibeat ECG-gated imaging
    a. Sequential acquisitions of volumes obtained from several ECG-gated consecutive heart cycles (from 2 to 6) that are stitched together to create a single volumetric

dataset

b. Improvements in spatial and temporal resolution

7. Real-time
   a. Used for full-volume acquisition
   b. Visualize small structures (e.g. aortic valve, masses)
   c. Record short-lived events such as saline contrast
   d. Image patients with irregular rhythm or dyspnea that prevent full-volume acquisition
   e. Monito interventional procedures
   f. Used during catheter interventions for atrial septal defects, ventricular septal defects and inflow valves
8. Live 3D color
   a. Color flow can be superimposed on a live 3D dataset to visualize blood flow in real time
   b. Temporal resolution is usually very low
9. 3D zoom
   a. Allows a focused real-time view of a structure of interest
   b. Assessment of atrial septal defects size and rim length
   c. Size and shape of VSD
10. 3D in congenital heart disease
    a. Used for pre-surgical planning, guidance of catheter interventions and functional assessment of the heart
    b. 3D TEE probes may be used for patients from neonate to adult
    c. Provides good spatial and temporal resolution
11. Clinical indications and imaging
    a. Use normal anatomic presentation in the presence of congenital heart disease
       i) As if a person is standing upright and facing you
    b. Atrial septal defects
       i) Visualized from right atrium
       ii) Landmarks include
          - Superior vena cava
          - Inferior vena cava
          - Ascending aorta
          - Fossa ovalis
          - Tricuspid valve
          - Coronary sinus
    c. Ventricular septal defects
       i) Visualized from RV aspect
       ii) RV outflow should be positioned upwards
       iii) RV apex to the right

iv) Landmarks include

- Tricuspid valve
- Moderator band
- Septomarginal trabeculation

d. Inflow valves

i) En-face from both ventricular and atrial side

ii) Diaphragmatic surface of the heart is shown at the lower part of the screen

iii) Can use 3D color and planimetry

iv) PLAX and PSAX

e. Outflow valves

i) Enface from outflow tract and great artery view

ii) PSAX and apical

iii) Can use 3D color planimetry

12. Modality comparison

a. Advantages of 3D/4D

b. Limitations of 3D/4D

c. Advantages of 2D

d. Limitations of 2D

Congenital Cardiac
Abbreviations

2D	Two dimensional
3D	Three dimensional

A

AI	Aortic insufficiency
AML	Anterior mitral leaflet
AO	Aorta
AoV	Aortic valve
AR	Aortic regurgitation
AS	Aortic stenosis
ASD	Atrial septal defect
ASH	Asymmetrical septal hypertrophy
ASO	Arterial switch operation
AVA	Aortic valve area
AVSD	Atrioventricular septal defect
AVV	Atrioventricular valve
AVVI	Atrioventricular valve index

B

BTTS	Blalock-Thomas-Taussig Shunt

C

CAD	Coronary artery disease
CCTGA	Congenitally corrected transposition of the great arteries
CHD	Congenital heart defect
CHF	Congestive heart failure
CI	Cardiac index
CO	Cardiac output
CS-ASD	Coronary sinus atrial septal defect
CW	Continuous wave

D

DCM	Dilated cardiomyopathy
DKS	Damus-Akye-Stansel
DORV	Double outlet right ventricle
DT	Deceleration time
D-TGA	Dextro transposition of the great arteries

Congenital Cardiac

E

EDV	End diastolic volume
EF	Ejection fraction
EFE	Endomyocardial fibroelastosis
EPSS	End point septal separation
EROA	Effective regurgitant orifice área
ESV	End systolic volume

F

FS	Fractional shortening

H

HCM	Hypertrophic cardiomyopathy
HLHS	Hypertrophic left heart syndrome

I

IAS	Interatrial septum
IVC	Inferior vena cava
IVCT	Isovolumic contraction time
IVRT	Isovolumic relaxation time
IVS	Interventricular septum

L

LA	Left atrium
LAA	Left atrial appendage
LAD	Left anterior descending
LBBB	Left bundle branch block
LCA	Left coronary artery
LCC	Left coronary cusp
LPA	Left pulmonary artery
LSVS	Left superior vena cava
LV	Left ventricle
LVEDP	Left ventricle end diastolic pressure
LVEDV	Left ventricle end diastolic volume
LVH	Left ventricular hypertrophy
LVOT	Left ventricular outflow tract
LVOTO	Left ventricular outflow tract obstruction

M

MAPCAs	Major Aortopulmonary collateral arteries
MPA	Main pulmonary artery
MR	Mitral regurgitation
MV	Mitral valve
MVA	Mitral valve area
MVP	Mitral valve prolapse

N

NCC	Non-coronary cusp

P

PA	Pulmonary artery
PAC	Premature atrial contraction
PAPVR	Partial anomalous pulmonary venous return
PDA	Patent Ductus Arteriosus
PFO	Patent foramen ovale
PHTN	Pulmonary hypertension
PLAX	Parasternal long axis
PSAX	Parasternal short axis
PVC	Premature ventricular contraction
PVR	Pulmonary vascular resistance
PW	Pulsed wave

R

RA	Right atrium
RBBB	Right bundle branch block
RCA	Right coronary artery
RCC	Right coronary cusp
RCM	Restrictive cardiomyopathy
RF	Regurgitant fraction
RIMP	Right ventricle index of myocardial performance
RPA	Right pulmonary artery
RV	Right ventricle
RVDCC	Right ventricle dependent coronary circulation
RVH	Right ventricular hypertrophy
RVOT	Right ventricular outflow tract
RVOTO	Right ventricular outflow tract obstruction

S

SA	Sinoatrial node
SAM	Systolic anterior motion
SOB	Shortness of breath
SpO_2	Oxygen saturation
STE	Speckle tacking echocardiography
SV	Stroke volume
SVC	Superior vena cava
SVD	Sinus venosum defect
SVT	Supraventricular tachycardia

T

TAPVR	Total anomalous pulmonary venous return
TDI	Tissue Doppler Imaging
TEE	Transesophageal echocardiogram
TOF	Tetralogy of Fallot
TR	Tricuspid regurgitation
TV	Tricuspid valve

V

Vef	Velocity of circumferential fiber shortening
VSD	Ventricular septal defect
VTI	Velocity time integral

W

WPW	Wolff-Parkinson-White

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Utilized References

1. Abuhamad A, Chaoui R. *A Practical Guide Fetal Echocardiography: Normal and Abnormal Hearts*, 4th ed. Lippincott Williams & Wilkens; 2021.

2. Anderson, B. *Echocardiography: The Normal Examination and Echocardiographic Measurements*, 3rd ed. MGA Graphics; 2016.

3. Anderson, B. *Sonographer's Guide to the Assessment of Heart Disease.* MGA Graphics; 2013.

4. ARDMS Pediatric Echocardiography Examination Content Outline. Accessed at PE-Examination-Content-Outline-V24.1-1pdf

5. Armstrong WF, Ryan T. *Feigenbaum's Echocardiography.* Wolters Kluwer; 2019

6. Bajraktari, G. *Echocardiography: In Specific Diseases.* IntechOpen; 2012.

7. Brown LM, Duffy CE, Mitchell C, Young L. A practical guide to pediatric coronary artery imaging with echocardiography. *Journal of the American Society of Echocardiography*. 2015;28(4):379-391.

8. CCI Registered Cardiac Sonographer (RCS) Exam Preparation. Accessed at https://cci-online.org/credentials/registered-cardiac-sonographer/

9. Cordes TM, O'Leary PW, Seward JB, Hagler DJ. Distinguishing right from left: A standardized technique for fetal echocardiography. *Journal of the American Society of Echocardiography*. 1994;7(1):47-53.

10. Crawford MH. *Current Diagnosis & Treatment. Cardiology,* 6th ed. McGraw-Hill Education; 2023.

11. Driscoll DJ. *Fundamentals of Pediatric Cardiology*. Wolters Kluwer Health; 2015.

12. Drose JA. *Fetal echocardiography*. Elsevier Health Sciences; 2010.

13. Eidem BW, Cetta F. *Echocardiography in Pediatric and Adult Congenital Heart Disease,* 3rd ed. Lippincott Williams & Wilkins; 2020.

14. Everett AD, Burns P, Lim DS, et al. *Illustrated Field Guide to Congenital Heart Disease and Repair*. Scientific Software Solutions; 2021.

15. Fuster V, Narula J, Vaishnava P, Leon MB, Callans DJ, Rumsfeld JS, Poppas A. *Fuster and Hurst's The Heart, 15th ed*. McGraw-Hill Education; 2022.

16. Gorman, et al., editors. The Sonographer Training HUB. The International Contrast Ultrasound Society, 2022, www. https://icus-society.org/education/sonographer-training-resource-hub/

17. Grubb NR, Newby DE. *Cardiology: An Illustrated Color Text.* Churchill Livingstone; 2005.

18. Harry MJ, Behrends T. *Essentials of Echocardiography: An Illustrative Guide*. Cardiac Ultrasound Consulting LLC; 2013.

19. Ho, SY, Rigby ML, Anderson RH. *Echocardiography in congenital Heart Disease Made Simple.* Imperial College Press; 2005.

20. Jaeggi E, Öhman A. Fetal and neonatal arrhythmias. *Clinics in Perinatology*. 2016;43(1):99-112.

21. Kucera F. *Atlas of Pediatric Echocardiography*. Elsevier; 2020.

22. Lai WW, Mertens LL, Cohen MS, Geva T. *Echocardiography in pediatric and congenital heart disease: From Fetus to Adult,* 3rd edition. John Wiley & Sons Inc; 2022.

23. Lang RM, Goldstein SA, Kronzon I, Khandheria BK, Saric M, Mor-Avi V. *ASE's Comprehensive Echocardiography*, 3rd ed. Philadelphia: Elsevier; 2022.

24. Lockwood CJ, Moore T MD, Copel J MD, Silver RM MD, Resnik R MD. *Creasy and Resnik's Maternal-Fetal Medicine: Principles and Practice*. Elsevier; 2022.

25. Lyshchik A. *Fundamentals of CEUS*. Elsevier; 2019.

26. McLaughlin ES, Schlosser BA, Border WL. Fetal diagnostics and fetal intervention. *Clinics in Perinatology*. 2016;43(1):23-38.

27. Moore KL, Persaud TVN, Torchia MG. *Before We Are Born: Essentials of Embryology and Birth Defects*. Saunders; 2019.

28. Norton ME, Callen PW. *Callen's ultrasonography in Obstetrics and Gynecology.* Saunders; 2016.

29. Oh JK, Kane GC, Seward JB, Tajik AJ. *The Echo Manual,* 4th ed. Lippincott-Raven; 2019.

30. Otto CM, Bonow, RO. *Valvular Heart Disease: A Companion to Braunwald's Heart Disease,* 4th ed. Elsevier Saunders; 2014.

31. Otto C. *Textbook of Clinical Echocardiography,* 7th ed. Elsevier; 2024.

32. Palma R. *Echocardiographer's Pocket Reference,* 5th ed. Arizona Heart Institute Foundation; 2020.

33. Pappano AJ, Wier WG. *Cardiovascular physiology*, 11th ed. Elsevier Health Sciences; 2018.

34. D'Andrea A, Radmilovic J, D'Amato A, et al. Echocardiography in athletes in primary prevention of sudden death. *Journal of Cardiovascular Echography*. 2019;29(4):139-148.

35. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy. *European Heart Journal.* 2018;39(34):3165-3241.

36. Rumack CM, Levin D. *Diagnostic Ultrasound*. Elsevier; 2018.

37. Rychik J, Tian Z. *Fetal Cardiovascular Imaging: A Disease-based Approach.* Saunders; 2011.

38. Sanapo L, Moon-Grady AJ, Donofrio MT. Perinatal and delivery management of infants with congenital heart disease. *Clinics in Perinatology*. 2016;43(1):55-71.

39. Snider RA, Serwer GA, Ritter SB. *Echocardiography in Pediatric Heart Disease,* 2nd ed. Mosby; 1997.

40. Van Hagen IM, Roos-Hesselink JW. Pregnancy in congenital heart disease: risk prediction and counselling. *Heart*. 2020;106(23):1853-1861.

41. Wilkins I, Hogge WA, Hill LM, Cohlan B. *Sanders' Structural Fetal Abnormalities,* 3rd ed. McGraw-Hill Education / Medical; 2016.

42. Yim D, Burgner D, Cheung M. Echocardiography in Kawasaki disease. IntechOpen. January 18, 2012. Accessed June 3, 2025. https://www.intechopen.com/chapters/26295.

Congenital Cardiac
ASE Guidelines References

1. Zhogbi WA, Jone PN, et al. Guidelines for the Evaluation of Prosthetic Valve Function With Cardiovascular Imaging. *Journal of the American Society of Echocardiography*. 2024;37:2-63.

2. Lopez L, Saurers DL, Barker PCA, et al. Guidelines for performing a Comprehensive Pediatric transthoracic echocardiogram: Recommendations from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2024;37(2):119-170.

3. McNamara PJ, Jain A, El-Khuffash A, et al. Guidelines and Recommendations for Targeted Neonatal Echocardiography and Cardiac Point-of-Care Ultrasound in the Neonatal Intensive Care Unit: An Update from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2024;37(2):171-215.

4. Mertens L, Singh G, Armenian S, et al. Multimodality imaging for cardiac surveillance of cancer treatment in children: Recommendations from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2023;36(12):1227-1253.

5. Moon-Grady AJ, Donofrio MT, Gelehrter S, et al. Guidelines and Recommendations for Performance of the Fetal Echocardiogram: An Update from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2023;36(7):679-723.

6. Lu JC, Riley A, Conlon T, et al. Recommendations for Cardiac Point-of-Care Ultrasound in Children: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2023;36(3):265-277.

7. Pandian NG, Kim JK, et al. Recommendations for the Use of Echocardiography in the Evaluation of Rheumatic Heart Disease: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2023;36:3-28.)

8. Nagueh SF, Phelan D, et al. Recommendations for Multimodality Cardiovascular Imaging of Patients with Hypertrophic Cardiomyopathy. *Journal of the American Society of Echocardiography.* 2022;35:533-69.

9. Pellikka PA, Arruda-Olson A, et al. Guidelines for Performance, Interpretation, and Application of Stress Echocardiography in Ischemic Heart Disease. *Journal of the American Society of Echocardiography.* 2020;33:1-41.

10. Zoghbi WA, Asch FM, et al. Guidelines for the Evaluation of Valvular Regurgitation After Percutaneous Valve Repair or Replacement. *Journal of the American Society of Echocardiography.* 2019;32:431-475.

11. Wasserman MA, Shea E, Cassidy C, et al. Recommendations for the adult cardiac sonographer performing echocardiography to screen for critical congenital heart disease in the newborn: from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2021;34(3):207-222.

12. Baggish AL, Battle RW, Beaver TA, et al. Recommendations on the Use of Multimodality Cardiovascular Imaging in Young Adult Competitive Athletes: A Report from the American Society of Echocardiography in Collaboration with the Society of Cardiovascular Computed Tomography and the Society for Cardiovascular Magnetic Resonance. *Journal of the American Society of Echocardiography.* 2020;33(5):523-549.

13. Frommelt P, Lopez L, Dimas VV, et al. Recommendations for Multimodality Assessment of Congenital Coronary Anomalies: A Guide from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2020;33(3):259-294.

14. Mitchell C, Rahko PS, et al. Guidelines for Performing a Comprehensive Transthoracic Echocardiographic Examination in Adults: Recommendations from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2019;32:1-64.

15. Porter TR, Mulvagh SL, et al. Clinical Applications of Ultrasonic Enhancing Agents in Echocardiography: 2018 American Society of Echocardiography Guidelines Update. *Journal of the American Society of Echocardiography.* 2018;31:241-274.

16. Zoghbi WA, Adams D, et al. Recommendations for Noninvasive Evaluation of Native Valvular Regurgitation. *Journal of the American Society of Echocardiography.* 2017;30:303-371.

17. Baumgartner H, Hung J, et al. Recommendations on the Echocardiographic Assessment of Aortic Valve Stenosis: A Focused Update. *Journal of the American Society of Echocardiography.* 2017;30:372-392.

18. Lancellotti P, Pellikka PA, et al. The Clinical Use of Stress Echocardiography in Non-Ischaemic Heart Disease. *Journal of the American Society of Echocardiography.* 2017;30:101-138.

19. Nagueh SF, Smiseth OA, et al. Recommendations for the Evaluation of Left Ventricular Diastolic Function by Echocardiography. *Journal of the American Society of Echocardiography.* 2016;29:277-314.

20. Simpson J, Lopez L, Acar P, et al. Three-dimensional Echocardiography in Congenital Heart Disease: An Expert Consensus Document from the European Association of Cardiovascular Imaging and the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2016;30(1):1-27.

21. Puchalski MD, Lui GK, Miller-Hance WC, et al. Guidelines for performing a comprehensive transesophageal echocardiographic. *Journal of the American Society of Echocardiography.* 2018;32(2):173-215.

22. Cohen MS, Eidem BW, Cetta F, et al. Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography.* 2016;29(7):571-621.

23. Silvestry FE, Cohen MS, Armsby LB, et al. Guidelines for the Echocardiographic Assessment of Atrial Septal Defect and Patent Foramen Ovale: from the American Society of Echocardiography and Society for Cardiac Angiography and Interventions. *Journal of the American Society of Echocardiography.* 2015;28(8):910-958.

24. Lang RM, Badano LP, Mor-Avi V, et al. Recommendations for Cardiac Chamber Quantification by Echocardiography in Adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *Journal of the American Society of Echocardiography.* 2015;28(1):1-39.

25. Campbell RM, Douglas PS, Eidem BW, Lai WW, Lopez L, Sachdeva R. ACC/AAP/AHA/ASE/HRS/SCAI/SCCT/SCMR/SOPE 2014 Appropriate Use Criteria for initial transthoracic echocardiography in outpatient pediatric cardiology. *Journal of the American College of Cardiology.* 2014;64(19):2039-2060.

26. McIlwain EF, Coon PD, Einstein AJ, et al. Radiation Safety for the Cardiac Sonographer: recommendations of the Radiation Safety Writing Group for the Council on Cardiovascular Sonography of the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2014;27(8):811-6.

27. Porter TR, Abdelmoneim S, Belcik JT, et al. Guidelines for the Cardiac Sonographer in the Performance of Contrast Echocardiography: a focused update from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2014;27(8):797-810.

28. Klein AL, Abbara S, Agler DA, et al. American Society of Echocardiography Clinical Recommendations for Multimodality Cardiovascular Imaging Of Patients With Pericardial Disease: endorsed by the Society for Cardiovascular Magnetic Resonance and Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography.* 2013;26(9):965-1012.

29. Reeves ST, Finely AC, Skubas NJ, et al. Basic Perioperative Transesophageal Echocardiography Examination: A Consensus Statement of the American Society of Echocardiography and the Society of

Cardiovascular Anesthesiologists. *Journal of the American Society of Echocardiography.* 2013;26(5):443-56.

30. Lang RM, Badano LP, Tsang W, et al. EAE/ASE Recommendations for Image Acquisition and Display using Three-dimensional Echocardiography. *Journal of the American Society of Echocardiography.* 2012;25(1):3-46.

31. Valente AM, Cook S, Festa P, et al. Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography.* 2014;27(2):111-141.

32. Zamorano JL, Badano LP, Bruce C, et al. EAE/ASE Recommendations for the Use of Echocardiography in New Transcatheter Interventions for Valvular Heart Disease. *Journal of the American Society of Echocardiography.* 2011;24(9):937-65.

33. Rudski LG, Lai WW, Afilalo J, et al. Guidelines for the Echocardiographic Assessment of the Right Heart in Adults. *Journal of the American Society of Echocardiography.* 2010;23(7):685-713.

34. Douglas, P, Garcia, M, Haines, D. et al. ACCF/ASE/AHA/ASNC/HFSA/HRS/SCAI/SCCM/SCCT/SCMR 2011 Appropriate Use Criteria for Echocardiography: A Report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, American Society of Echocardiography, American Heart Association, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, Society of Critical Care Medicine, Society of Cardiovascular Computed Tomography, and Society for Cardiovascular Magnetic Resonance Endorsed by the American College of Chest Physicians. Journal of the American College of Cardiology. 2011; 57(9):1126–1166.

