

THE AMERICAN BOARD OF PEDIATRICS®

# CONTENT OUTLINE

## Pediatric Cardiology

**Subspecialty In-training, Certification, and  
Maintenance of Certification Examinations**

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## **INTRODUCTION**

This document was prepared by the American Board of Pediatrics Subboard of Pediatric Cardiology for the purpose of developing in-training, certification, and maintenance of certification examinations. The outline defines the body of knowledge from which the Subboard samples to prepare its examinations. The content specification statements located under each category of the outline are used by item writers to develop questions for the examinations; they broadly address the specific elements of knowledge within each section of the outline.

## Pediatric Cardiology

Each Pediatric Cardiology exam is built to the same specifications, also known as the blueprint. This blueprint is used to ensure that, for the initial certification and in-training exams, each exam measures the same depth and breadth of content knowledge. Similarly, the blueprint ensures that the same is true for each Maintenance of Certification exam form. The table below shows the percentage of questions from each of the content domains that will appear on an exam. Please note that the percentages are approximate; actual content may vary.

	Content Categories	Initial Certification and In-Training	Maintenance of Certification (MOC)
1.	Cardiovascular Structure, Development, and Function	7%	6%
2.	Pharmacology	6%	6%
3.	Cardiovascular Examination, Principles, and Application of Cardiac Diagnostics	9%	10%
4.	Office-Based Cardiac Problems	7%	10%
5.	Left-to-Right Shunts	6%	6%
6.	Right-to-Left Shunts	6%	5%
7.	Single Ventricular Lesions	6%	6%
8.	Structural, Valvar, and Obstructive Lesions	6%	6%
9.	Congenital Abnormalities of the Great Arteries and Aorta	5%	5%
10.	Systemic and Pulmonary Venous Abnormalities and Situs Abnormalities	4%	4%
11.	Disorders of the Myocardium, Pericardium, Endocardium, and Vasculature	4%	4%
12.	Heart Function and Disease in the Fetus and Newborn	5%	5%
13.	Intensive Care Management of Patients with Congenital Heart Disease	5%	4%
14.	Arrhythmias	6%	6%
15.	Acquired Forms of Cardiac Disease	4%	4%
16.	Genetic Disorders and Syndromes of the Cardiovascular System	4%	4%
17.	Congenital Heart Disease in the Adolescent and Adult	5%	6%
18.	Core Knowledge in Scholarly Activities	5%	3%

# Cardiology

## 1. Cardiopulmonary Structure, Development, and Function

### A. Cardiovascular anatomy and ultrastructure

#### 1. Gross cardiovascular anatomy

- a. Understand the principles of segmental anatomy
- b. Know the normal anatomy of the systemic veins and important variations
- c. Know the normal anatomy of the atria, atrial septum, appendages, and relationship to thoracic structures
- d. Know the anatomic features of the atrioventricular valves and atrioventricular orientation (concordance/discordance)
- e. Know the normal anatomical features of the right ventricle, left ventricle, and the interventricular septum, and ventriculoarterial orientation (concordance/discordance)
- f. Know the anatomic features of semilunar valves, including spatial relationships to each other and the ventricular outlets
- g. Know the anatomical features of the pulmonary arteries, pulmonary vasculature, and orientation to thoracic structures (eg, bronchi)
- h. Know the normal structure and anatomic relationships of pulmonary veins
- i. Know the normal anatomic features of the coronary arteries
- j. Know the normal anatomy of the aorta and its orientation to thoracic structures
- k. Know the normal anatomy of the systemic arteries and important variations
- l. Understand the relationship between the left recurrent laryngeal nerve and structures within the thorax

#### 2. Ultrastructure

- a. Know the ultrastructure of cardiac myocyte along with developmental aspects of the myofibrils, contractile proteins, transverse tubular system, sarcoplasmic reticulum, intercalated disc, nucleus and perinuclear region, mitochondria, and glycogen stores

### B. Normal embryology and development of myocardial structure and function

1. Know the important components (tissues) of the primitive cardiac tube and the origins of cardiac segments including origin of the primary and secondary heart fields
2. Understand looping of the primitive cardiac tube under normal and abnormal conditions
3. Know the normal development of the peripheral vasculature
4. Describe normal embryologic sequence in the development of pulmonary veins, including the fate of the common pulmonary vein
5. Know the normal development of the bulbus cordis and truncus arteriosus
6. Know the embryologic basis for atrioventricular connections
7. Know the development of the six aortic arches (eg, sequence, timing, regressions)
8. Know the normal development of the various cardiovascular structures (eg, semilunar valves, aortic pulmonary septum, division of truncus)
9. Understand the contribution of neural crest tissue to normal conotruncal development
10. Understand the molecular regulation of cardiac determination and differentiation

11. Identify the molecular pathways that determine visceral situs
  12. Know the origins of the epicardium and its importance for coronary, valvar, and fibrous development of the heart
  13. Know the normal development of the conduction system
  14. Know the normal developmental sequence of the pulmonary vasculature
  15. Know the normal developmental sequence of the coronary vasculature
- C. Cardiovascular physiology
1. Embryo and fetus
    - a. Know the distribution of cardiac output to the various circulations during normal and abnormal conditions (eg, pulmonary, coronary, central nervous system, ventricular, peripheral vascular beds, placenta)
    - b. Understand how blood flow patterns in cardiovascular structures, including fetal structures, may be influenced by congenital anomalies
    - c. Understand regulation of circulation in the embryo and fetus
    - d. Understand the fetal circulation and its clinical correlates with findings on fetal echocardiography
  2. Postnatal circulation
    - a. Understand the physiologic aspects of postnatal heart rate, preload effects, afterload effects, and contractility, and how these factors modulate cardiac output
    - b. Know the postnatal changes in pulmonary and systemic circulations after birth and how they may be influenced by congenital defects (eg, left-to-right shunts, hypoxemia, obstructive lesions)
  3. Cardiac mechanics
    - a. Understand the interaction of contractile and regulatory protein in determining sarcomere function
    - b. Understand the role of calcium ions in the contractile process and dynamics of calcium ion movement/storage
    - c. Understand the process of excitation-contraction coupling
    - d. Understand the concepts and variation of length-tension relationships
    - e. Understand ventricular pressure-volume function loops
    - f. Understand the concept of interventricular interaction
    - g. Understand the role of neural-humoral factors on control of the cardiovascular system
    - h. Understand the role of the thyroid in modulating control of the cardiovascular system
    - i. Understand the role of baroreceptors in control of the cardiovascular system
    - j. Understand the role of chemoreceptors in control of the cardiovascular system
    - k. Understand cellular responses and their interactions with drugs, blood gases, and pH
    - l. Know the molecular events occurring during the cardiac cycle
    - m. Understand the role of the pericardium on impairment of cardiac function
    - n. Understand myocardial metabolism and the impact of hormonal influences, hypoxia, ischemia, and age
    - o. Know how hemodynamic loads affect myocardial gene expression
    - p. Recognize the acute effects and compensatory responses of ventricular dilatation and hypertrophy on cardiac function

- q. Understand the atrial contribution of ventricular function
  - r. Know the age-related differences in function of sarcolemma and sarcoplasmic reticulum
  - s. Know the effects of acidemia and hypoxemia on contractility
  - t. Recognize factors involved in the myocardial oxygen supply-demand ratio
  - u. Know how hemodynamic loads affect vascular gene expression
  - v. Understand cardiac receptor function and its response to drugs, blood gases, and pH
  - w. Understand the determinants of contractions of isolated cardiac myocytes and papillary muscle
4. Vascular physiology
- a. Understand the concept of autoregulation of regional blood flow, including age-related differences
  - b. Know the role of inflammatory mediators and of endothelium in control of regional circulation
  - c. Understand the mechanisms that regulate coronary circulation, including exercise and hypoxemia
  - d. Understand the mechanisms that regulate cerebral circulation, including pH, PCO<sub>2</sub>, PO<sub>2</sub>, and perfusion
  - e. Understand the influence of pH, PO<sub>2</sub>, and PCO<sub>2</sub> on pulmonary circulation
  - f. Understand the role of endothelium on pulmonary circulation
  - g. Understand the mechanisms involved in vasodilation in pulmonary and systemic vascular beds
  - h. Identify the factors influencing tone in the pulmonary vascular bed
  - i. Understand how vascular smooth muscle is maintained in a dilated state
  - j. Understand the Poiseuille equation and the effect of changing the caliber of the resistance vessels
  - k. Know the effects of asphyxia on regional circulatory functions, particularly heart, brain, kidneys, and lung
  - l. Know the effects of changes in renal blood flow on renal function and body fluids
  - m. Understand the mechanisms that regulate skeletal muscle blood flow during exercise
- D. Conduction system, including electrophysiology
- 1. Developmental aspects
    - a. Recognize the age-dependent nature of structure and function of the conduction system
  - 2. Anatomic aspects
    - a. Know anatomic basis and features of the conduction system
    - b. Know the conduction system abnormalities associated with cardiac anomalies
  - 3. Electrophysiologic aspects
    - a. Understand the structure and function of ion channels in myocardial tissue
    - b. Understand the components of the myocardial cell action potential
    - c. Understand the ionic basis of cardiac automaticity
    - d. Know the electrophysiologic characteristics of the atrial, atrioventricular, and ventricular conduction system
- E. Respiratory physiology and ventilation

1. Structure
  - a. Understand the age-related (including prematurity) effects on airway physiology, including airway resistance and ventilation pattern
  - b. Recognize the effects of upper airway obstruction on cardiopulmonary physiology
  - c. Recognize effects of large airway obstruction on cardiopulmonary physiology
  - d. Recognize effects of cardiomegaly and pulmonary vascular anatomy on large airway function
  - e. Know the pathologic changes that occur in the small airways in various cardiopulmonary conditions
  - f. Recognize potential effects of cardiomegaly and pulmonary vasculature on small airway function
  - g. Know the pathologic changes that occur in the alveoli in various cardiopulmonary conditions, including congestive heart failure and alterations in pulmonary blood flow
2. Ventilation
  - a. Identify and know importance of variations in pulmonary pressure-volume relationships
  - b. Understand the effects of FIO<sub>2</sub> on nitrogen clearance, regional blood flow, and arterial blood gas tensions
  - c. Know the importance of normal lung inflation and deflation on cardiovascular physiology
  - d. Recognize the influence of cardiovascular disease on lung volume, tidal volume, and respiratory rate
3. Oxygen delivery
  - a. Understand the determinants of gas transfer from the airway to arterial blood
  - b. Know factors influencing the amount of dissolved oxygen in blood
  - c. Understand the differences between adult and fetal hemoglobin
  - d. Understand the effects of a change in arterial oxygen tension in the oxyhemoglobin dissociation curve, and mixed venous oxygen content
  - e. Apply the principles of ventilation-perfusion balance and mismatch in the evaluation of a patient with cardiovascular disease
4. Acid-base balance
  - a. Know how to recognize acid-base abnormalities, including respiratory acidosis and alkalosis and metabolic acidosis and alkalosis
  - b. Identify the causes of acid-base abnormalities, including respiratory acidosis and alkalosis and metabolic acidosis and alkalosis
  - c. Know methods for correction of acid-base abnormalities, including respiratory alkalosis and acidosis and metabolic acidosis and alkalosis
5. Blood gas exchange
  - a. Understand the effects of ischemia on O<sub>2</sub> and CO<sub>2</sub> exchange between blood and tissue
  - b. Understand the effects of pH, hypoxemia, and hypercarbia on gas exchange between blood and tissue
  - c. Know the effects of hypoxemia on ventilatory function and tissue metabolism
6. Mechanism of breathing
  - a. Understand the possible physiologic effects of chest wall abnormality

- b. Understand the effects of dynamic compression of airways
- c. Recognize the effect of respiratory effort on caloric consumption
- 7. Respiratory physiology at altitude
  - a. Recognize the effects of acute and chronic exposure to high altitude on oxygen delivery
  - b. Know the effects of altitude on cardiovascular function
- 8. Effects of mechanical ventilation on CV performance
  - a. Understand the effects of positive end-expiratory pressure and inspiratory pressures on cardiac output and their influence on right and left ventricular function
  - b. Understand appropriate use of mechanical ventilation
  - c. Know the general principles of how the mode of ventilation affects cardiac output

## 2. Pharmacology

- A. Pharmacologic basis for therapy of cardiovascular disease
  - 1. Understand the principles of pharmacokinetics, including drug absorption, distribution, biotransformation, and excretion
  - 2. Understand the principles of pharmacodynamics, including the loci of drug actions, role of receptors, and dose-response relationships
  - 3. Understand drug-drug interactions, adverse reactions, and off-target effects
  - 4. Understand the principles of maternal-fetal pharmacodynamics and pharmacokinetics
  - 5. Understand the basic principles of developmental pharmacology
  - 6. Understand the role of pharmacogenomics in drug response
  - 7. Understand the principles involved in clinical trials and drug regulation and development
- B. Antiarrhythmic drugs
  - 1. Class I antiarrhythmic drugs
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of Class I antiarrhythmic drugs
    - b. Know the mechanism of action for Class I antiarrhythmic drugs
    - c. Identify indications for Class I antiarrhythmic drugs
    - d. Identify contraindications for Class I antiarrhythmic drugs
    - e. Plan therapy including appropriate dose and monitoring for Class I antiarrhythmic drugs
    - f. Recognize the drug adverse effects and toxicity of Class I antiarrhythmic drugs
    - g. Recognize potential drug-drug interactions for Class I antiarrhythmic drugs
  - 2. Class II antiarrhythmic drugs
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of Class II antiarrhythmic drugs
    - b. Know the mechanism of action for Class II antiarrhythmic drugs
    - c. Identify Indications for Class II antiarrhythmic drugs
    - d. Identify contraindications for Class II antiarrhythmic drugs
    - e. Plan therapy including appropriate dose and monitoring for Class II antiarrhythmic drugs
    - f. Recognize the drug adverse effects and toxicity of Class II antiarrhythmic drugs
    - g. Recognize potential drug-drug interactions for Class II antiarrhythmic drugs
  - 3. Class III drugs



- a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of Class III antiarrhythmic drugs
  - b. Know the mechanism of action for Class III antiarrhythmic drugs
  - c. Identify indications for Class III antiarrhythmic drugs
  - d. Identify contraindications for Class III antiarrhythmic drugs
  - e. Plan therapy including appropriate dose and monitoring for Class III antiarrhythmic drugs
  - f. Recognize the drug adverse effects and toxicity of Class III antiarrhythmics
  - g. Recognize potential drug-drug interactions for Class III antiarrhythmic drugs
  4. Class IV antiarrhythmic drugs
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of Class IV antiarrhythmic drugs
    - b. Know the mechanism of action for Class IV antiarrhythmic drugs
    - c. Identify indications for Class IV antiarrhythmic drugs
    - d. Identify contraindications for Class IV antiarrhythmic drugs
    - e. Plan therapy including appropriate dose and monitoring for Class IV antiarrhythmic drugs
    - f. Recognize the drug adverse effects and toxicity of Class IV antiarrhythmics
    - g. Recognize potential drug-drug interactions for Class IV antiarrhythmic drugs
  5. Adenosine
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of adenosine
    - b. Know the mechanism of action for adenosine
    - c. Identify indications for adenosine
    - d. Identify contraindications for adenosine
    - e. Plan therapy including appropriate dose and monitoring for adenosine
    - f. Recognize the drug adverse effects and toxicity of adenosine
    - g. Recognize potential drug-drug interactions for adenosine
- C. Inotropic agents
1. Digitalis and ATPase inhibitors
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of digitalis and ATPase inhibitors
    - b. Know the mechanism of action for digitalis and ATPase inhibitors
    - c. Identify indications for digitalis and ATPase inhibitors
    - d. Identify contraindications for digitalis and ATPase inhibitors
    - e. Plan therapy including appropriate dose and monitoring for digitalis and ATPase inhibitors
    - f. Recognize the drug adverse effects and toxicity of digitalis and ATPase inhibitors
    - g. Recognize potential drug-drug interactions for digitalis and ATPase inhibitors
  2. Catecholamines
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of catecholamines
    - b. Know the mechanism of action for catecholamines
    - c. Identify indications for catecholamines
    - d. Identify contraindications for catecholamines
    - e. Plan therapy including appropriate dose and monitoring for catecholamines

- f. Recognize the drug adverse effects and toxicity of catecholamines
- g. Recognize potential drug-drug interactions for catecholamines
- 3. Phosphodiesterase inhibitors
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of phosphodiesterase inhibitors
  - b. Know the mechanism of action for phosphodiesterase inhibitors
  - c. Identify indications for phosphodiesterase inhibitors
  - d. Identify contraindications for phosphodiesterase inhibitors
  - e. Plan therapy including appropriate dose and monitoring for phosphodiesterase inhibitors
  - f. Recognize the drug adverse effects and toxicity of phosphodiesterase inhibitors
  - g. Recognize potential drug-drug interactions for phosphodiesterase inhibitors
- 4. Calcium sensitizers
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of calcium sensitizers
  - b. Know the mechanism of action for calcium sensitizers
  - c. Identify indications for calcium sensitizers
  - d. Identify contraindications for calcium sensitizers
  - e. Plan therapy including appropriate dose and monitoring for calcium sensitizers
  - f. Recognize the drug adverse effects and toxicity of calcium sensitizers
  - g. Recognize potential drug-drug interactions for calcium sensitizers
- D. Systemic vasoactive agents
  - 1. Nitrates
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of nitrates
    - b. Know the mechanism of action for nitrates
    - c. Identify indications for nitrates
    - d. Identify contraindications for nitrates
    - e. Plan therapy including appropriate dose and monitoring for nitrates
    - f. Recognize the drug adverse effects and toxicity of nitrates
    - g. Recognize potential drug-drug interactions for nitrates
  - 2. Calcium channel antagonists
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of calcium channel antagonists
    - b. Know the mechanism of action for calcium channel antagonists
    - c. Identify indications for calcium channel antagonists
    - d. Identify contraindications for calcium channel antagonists
    - e. Plan therapy including appropriate dose and monitoring for calcium channel antagonists
    - f. Recognize the drug adverse effects and toxicity of calcium channel antagonists
    - g. Recognize potential drug-drug interactions for calcium channel antagonists
  - 3. Angiotensin-converting enzyme (ACE) inhibitors
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of ACE inhibitors
    - b. Know the mechanism of action for ACE Inhibitors
    - c. Identify indications for ACE inhibitors

- d. Identify contraindications for ACE inhibitors
- e. Plan therapy including appropriate dose and monitoring for ACE inhibitors
- f. Recognize the drug adverse effects and toxicity of ACE inhibitors
- g. Recognize potential drug-drug interactions for ACE inhibitors
- 4. Beta-adrenergic blocking drugs
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of  $\beta$ -adrenergic blocking drugs
  - b. Know the mechanism of action for  $\beta$ -adrenergic blocking drugs
  - c. Identify indications for  $\beta$ -adrenergic blocking drugs
  - d. Identify contraindications for  $\beta$ -adrenergic blocking drugs
  - e. Plan therapy including appropriate dose and monitoring for  $\beta$ -adrenergic blocking drugs
  - f. Recognize the drug adverse effects and toxicity of  $\beta$ -adrenergic blocking drugs
  - g. Recognize potential drug-drug interactions for  $\beta$ -adrenergic blocking drugs
- 5. Angiotensin-receptor blockers (ARBs)
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of ARBs
  - b. Know the mechanism of action for ARBs
  - c. Identify indications for ARBs
  - d. Identify contraindications for ARBs
  - e. Plan therapy including appropriate dose and monitoring for ARBs
  - f. Recognize the drug adverse effects and toxicity of ARBs
  - g. Recognize potential drug-drug interactions for ARBs
- 6. Fenoldopam
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of fenoldopam
  - b. Know the mechanism of action for fenoldopam
  - c. Identify indications for fenoldopam
  - d. Identify contraindications for fenoldopam
  - e. Plan therapy including appropriate dose and monitoring for fenoldopam
  - f. Recognize the drug adverse effects and toxicity of fenoldopam
  - g. Recognize potential drug-drug interactions for fenoldopam
- 7. Vasopressin
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of vasopressin
  - b. Know the mechanism of action for vasopressin
  - c. Identify indications for vasopressin
  - d. Identify contraindications for vasopressin
  - e. Plan therapy including appropriate dose and monitoring for vasopressin
  - f. Recognize the drug adverse effects and toxicity of vasopressin
  - g. Recognize potential drug-drug interactions for vasopressin
- E. Pulmonary vasoactive agents
  - 1. Prostacyclin, epoprostenol, sildenafil, bosentan, etc.
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of pulmonary vasodilators
    - b. Know the mechanism of action for pulmonary vasodilators

- c. Identify indications for pulmonary vasodilators
  - d. Identify contraindications for pulmonary vasodilators
  - e. Plan therapy including appropriate dose and monitoring for pulmonary vasodilators
  - f. Recognize the drug adverse effects and toxicity of pulmonary vasodilators
  - g. Recognize potential drug-drug interactions for pulmonary vasodilators
- F. Diuretics
- 1. Loop diuretics
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of loop diuretics
    - b. Know the mechanism of action for loop diuretics
    - c. Identify indications for loop diuretics
    - d. Identify contraindications for loop diuretics
    - e. Plan therapy including appropriate dose and monitoring for loop diuretics
    - f. Recognize the drug adverse effects and toxicity of loop diuretics
    - g. Recognize potential drug-drug interactions for loop diuretics
  - 2. Thiazides
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of thiazides
    - b. Know the mechanism of action for thiazides
    - c. Identify indications for thiazides
    - d. Identify contraindications for thiazides
    - e. Plan therapy including appropriate dose and monitoring for thiazides
    - f. Recognize the drug adverse effects and toxicity of thiazides
    - g. Recognize potential drug-drug interactions for thiazides
  - 3. Carbonic anhydrase inhibitors
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of carbonic anhydrase inhibitors
    - b. Know the mechanism of action for carbonic anhydrase inhibitors
    - c. Identify indications for carbonic anhydrase inhibitors
    - d. Identify contraindications for carbonic anhydrase inhibitors
    - e. Plan therapy including appropriate dose and monitoring for carbonic anhydrase inhibitors
    - f. Recognize the drug adverse effects and toxicity of carbonic anhydrase inhibitors
    - g. Recognize potential drug-drug interactions for carbonic anhydrase inhibitors
  - 4. Potassium-sparing diuretics
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of potassium-sparing diuretics
    - b. Know the mechanism of action for potassium-sparing diuretics
    - c. Identify indications for potassium-sparing diuretics
    - d. Identify contraindications for potassium-sparing diuretics
    - e. Plan therapy including appropriate dose and monitoring for potassium-sparing diuretics
    - f. Recognize the drug adverse effects and toxicity of potassium-sparing diuretics
    - g. Recognize potential drug-drug interactions for potassium-sparing diuretics
  - 5. B-type natriuretic peptides

- a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of b-type natriuretic peptides
  - b. Know the mechanism of action for b-type natriuretic peptides
  - c. Identify indications for b-type natriuretic peptides
  - d. Identify contraindications for b-type natriuretic peptides
  - e. Plan therapy including appropriate dose and monitoring for b-type natriuretic peptides
  - f. Recognize the drug adverse effects and toxicity of b-type natriuretic peptides
  - g. Recognize potential drug-drug interactions for b-type natriuretic peptides
- G. Anti-inflammatory agents
1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of anti-inflammatory agents
  2. Know the mechanism of action of anti-inflammatory agents
  3. Identify indications for anti-inflammatory agents
  4. Identify contraindications for anti-inflammatory agents
  5. Plan therapy including appropriate dose and monitoring for anti-inflammatory agents
  6. Recognize the drug adverse effects and toxicity of anti-inflammatory agents
  7. Recognize potential drug-drug interactions for anti-inflammatory agents
- H. Antiplatelet/anticoagulant therapy
1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of antithrombic/anticoagulant agents
  2. Know the mechanism of action for antithrombic/anticoagulant agents
  3. Identify indications for antithrombic/anticoagulant agents
  4. Identify contraindications for antithrombic/anticoagulant agents
  5. Plan therapy including appropriate dose and monitoring for antithrombic/anticoagulant agents
  6. Recognize the drug adverse effects and toxicity of antithrombic/anticoagulant agents
  7. Recognize potential drug-drug interactions for antithrombic/anticoagulant agents
- I. Lipid-lowering drugs
1. Bile acid sequestrants
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of lipid-reducing agents
    - b. Know the mechanism of action for lipid-reducing agents
    - c. Identify indications for lipid-reducing agents
    - d. Identify contraindications for lipid-reducing agents
    - e. Plan therapy including appropriate dose and monitoring for lipid-reducing agents
    - f. Recognize the drug adverse effects and toxicity of lipid-reducing agents
    - g. Recognize potential drug-drug interactions for lipid-reducing agents
  2. HMG CoA reductase inhibitors
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of HMG CoA reductase inhibitors
    - b. Know the mechanism of action for HMG CoA reductase inhibitors
    - c. Identify indications for HMG CoA reductase inhibitors
    - d. Identify contraindications for HMG CoA reductase inhibitors
    - e. Plan therapy including appropriate dose and monitoring for HMG CoA reductase inhibitors

- f. Recognize the drug adverse effects and toxicity of HMG CoA reductase inhibitors
- g. Recognize potential drug-drug interactions for HMG CoA reductase inhibitors
- 3. Glycoprotein IIb/IIIa inhibitors
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of glycoprotein IIb/IIIa inhibitors
  - b. Know the mechanism of action for glycoprotein IIb/IIIa inhibitors
  - c. Identify indications for glycoprotein IIb/IIIa inhibitors
  - d. Identify contraindications for glycoprotein IIb/IIIa inhibitors
  - e. Plan therapy including appropriate dose and monitoring for glycoprotein IIb/IIIa inhibitors
  - f. Recognize the drug adverse effects and toxicity of glycoprotein IIb/IIIa inhibitors
  - g. Recognize potential drug-drug interactions for glycoprotein IIb/IIIa inhibitors
- J. Ductal arteriosus drugs
  - 1. Prostaglandins
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of prostaglandin E (PGE)
    - b. Know the mechanism of action for PGE
    - c. Identify indications for PGE
    - d. Identify contraindications for PGE
    - e. Plan therapy including appropriate dose and monitoring for PGE
    - f. Recognize the drug adverse effects and toxicity of PGE
    - g. Recognize potential drug-drug interactions for PGE
  - 2. Indomethacin and ibuprofen
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of indomethacin/ibuprofen
    - b. Know the mechanism of action for indomethacin/ibuprofen
    - c. Identify indications for indomethacin/ibuprofen
    - d. Identify contraindications for indomethacin/ibuprofen
    - e. Plan therapy including appropriate dose and monitoring for indomethacin/ibuprofen
    - f. Recognize the drug adverse effects and toxicity of indomethacin/ibuprofen
    - g. Recognize potential drug-drug interactions for indomethacin/ibuprofen
- K. Specific sedative, hypnotic, and analgesic drugs
  - 1. Barbiturates
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of barbiturates
    - b. Know the mechanism of action of barbiturates
    - c. Identify indications for barbiturates
    - d. Identify contraindications for barbiturates
    - e. Plan therapy including appropriate dose and monitoring for barbiturates
    - f. Recognize the drug adverse effects and toxicity of barbiturates
    - g. Recognize potential drug-drug interactions for barbiturates
  - 2. Benzodiazepines and antagonists (eg, diazepam, midazolam, flumazenil)
    - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of benzodiazepines and antagonists
    - b. Know the mechanism of action for benzodiazepines and antagonists

- c. Identify indications for benzodiazepines and antagonists
- d. Identify contraindications for benzodiazepines and antagonists
- e. Plan therapy including appropriate dose and monitoring for benzodiazepines and antagonists
- f. Recognize the drug adverse effects and toxicity of benzodiazepines and antagonists
- g. Recognize potential drug-drug interactions for benzodiazepines and antagonists
- 3. Non-barbiturates/non-benzodiazepines (eg, etomidate, ketamine)
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of non-barbiturate/non-benzodiazepine agents
  - b. Know the mechanism of action for non-barbiturate/non-benzodiazepine agents
  - c. Identify indications for non-barbiturate/non-benzodiazepine agents
  - d. Identify contraindications for non-barbiturate/non-benzodiazepine agents
  - e. Plan therapy including appropriate dose and monitoring for non-barbiturate/non-benzodiazepine agents
  - f. Recognize the drug adverse effects and toxicity of non-barbiturate/non-benzodiazepine agents
  - g. Recognize potential drug-drug interactions for non-barbiturate/non-benzodiazepine agents
- 4. Opioids (eg, morphine, fentanyl, meperidine, sufentanil, methadone)
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of opioids
  - b. Know the mechanism of action for opioids
  - c. Identify indications for opioids
  - d. Identify contraindications for opioids
  - e. Plan therapy including appropriate dose and monitoring for opioids
  - f. Recognize the drug adverse effects and toxicity of opioids
  - g. Recognize potential drug-drug interactions for opioids
- 5. Psychotropic drugs
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of psychotropic drugs
  - b. Know the mechanism of action for psychotropic drugs
  - c. Identify indications for psychotropic drugs
  - d. Identify contraindications for psychotropic drugs
  - e. Plan therapy including appropriate dose and monitoring for psychotropic drugs
  - f. Recognize the drug adverse effects and toxicity of psychotropic drugs
  - g. Recognize potential drug-drug interactions for psychotropic drugs
- 6. Local anesthetics (eg, lidocaine, mepivacaine)
  - a. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of local anesthetics
  - b. Know the mechanism of action for local anesthetics
  - c. Identify indications for local anesthetics
  - d. Identify contraindications for local anesthetics
  - e. Plan therapy including appropriate dose and monitoring for local anesthetics
  - f. Recognize the drug adverse effects and toxicity of local anesthetics
  - g. Recognize potential drug-drug interactions for local anesthetics
- L. Anti-cholinergic drugs

1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of anti-cholinergic drugs
  2. Know the mechanism of action for anti-cholinergic drugs
  3. Identify indications for anti-cholinergic drugs
  4. Identify contraindications for anti-cholinergic drugs
  5. Plan therapy including appropriate dose and monitoring for anti-cholinergic drugs
  6. Recognize the drug adverse effects and toxicity of anti-cholinergic drugs
  7. Recognize potential drug-drug interactions for anti-cholinergic drugs
- M. Neuromuscular blocking agents
1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of neuromuscular blocking drugs
  2. Know the mechanism of action for neuromuscular blocking drugs
  3. Identify indications for neuromuscular blocking drugs
  4. Identify contraindications for neuromuscular blocking drugs
  5. Plan therapy including appropriate dose and monitoring for neuromuscular blocking drugs
  6. Recognize the drug adverse effects and toxicity of neuromuscular blocking drugs
  7. Recognize potential drug-drug interactions for neuromuscular blocking drugs
- N. Inhalation anesthetics
1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of inhalation anesthetics
  2. Know the mechanism of action for inhalation anesthetics
  3. Identify indications for inhalation anesthetics
  4. Identify contraindications for inhalation anesthetics
  5. Plan therapy including appropriate dose and monitoring for inhalation anesthetics
  6. Recognize the drug adverse effects and toxicity of inhalation anesthetics
  7. Recognize potential drug-drug interactions for inhalation anesthetics
- O. Antireflux drugs
1. Antireflux medications
    - a. Understand the cardiovascular effects of antireflux drugs, including contraindications and interactions with other drugs
    - b. Know the mechanism of action for antireflux drugs
    - c. Identify indications for antireflux drugs
    - d. Identify contraindications for antireflux drugs
    - e. Plan therapy including appropriate dose and monitoring for antireflux drugs
    - f. Recognize the drug adverse effects and toxicity of antireflux drugs
    - g. Recognize potential drug-drug interactions for antireflux drugs
- P. Enzyme replacement therapy for Pompe, Fabry, Hurler
1. Know the pharmacologic effects (pharmacokinetics and pharmacodynamics) of enzyme replacement therapy
  2. Know the mechanism of action for enzyme replacement therapy
  3. Identify indications for enzyme replacement therapy
  4. Identify contraindications for enzyme replacement therapy
  5. Plan therapy including appropriate dose and monitoring for enzyme replacement therapy
  6. Recognize the drug adverse effects and toxicity of enzyme replacement therapy



7. Recognize potential drug-drug interactions for enzyme replacement therapy
- Q. Cardiovascular effects of drug abuse
  1. Recognize and manage the cardiovascular effects of drug abuse (eg, cocaine, diet pills, hallucinogens, inhalants, stimulants, anabolic steroids)
3. **Cardiovascular Examination, Principles, and Application of Cardiac Diagnostics**
  - A. Physical examination
    1. Heart rate
      - a. Know the age-related normal measurements of heart rate associated with disease states and medications
      - b. Understand alterations in heart rate and types of rhythm associated with disease states and medications
    2. Vasculature including arterial and venous pulses, venous congestion, and blood pressure
      - a. Know significance of differences in pulse amplitude between extremities
      - b. Understand the physiology of paradoxical pulse
      - c. Understand the significance of a widened pulse pressure
      - d. Know the age-related changes in blood pressure values
      - e. Know the different methods of determination of blood pressure and the potential associated artifacts
      - f. Understand the physiologic events related to the jugular venous pulse and causes for variation
      - g. Know the principles that underlie the assessment of perfusion (eg, temperature, capillary refilling, color)
      - h. Recognize the clinical signs of systemic venous congestion and know the significance of peripheral edema in patients with cardiac disease
    3. Murmurs
      - a. Understand the physical basis of murmurs, thrills, and ventricular heaves
      - b. Know characteristics of normal and abnormal heart sounds with respect to physiologic events and timing in the cardiac cycle
      - c. Recognize the characteristics of the various types of functional ("innocent") murmurs
      - d. Understand the significance of localization and transmission of cardiac murmurs
      - e. Know how to interpret extracardiac bruits
      - f. Understand the significance of friction rubs
      - g. Know the various characteristics of pathologic murmurs, clicks, and cardiac sounds.
      - h. Interpret clinical physical examination data influenced by cardiac and body position
    4. Respiratory pattern
      - a. Recognize the clinical signs of pulmonary congestion
      - b. Know the clinical significance of abnormal respiratory patterns (eg, tachypnea, hyperpnea, stridor, grunting, retractions, wheezing)
    5. Cyanosis and other skin manifestations of cardiac disease
      - a. Know the cardiac and noncardiac causes of central cyanosis
      - b. Understand the physiologic significance of central versus peripheral cyanosis
      - c. Know the significance of jaundice in a cardiac patient

- d. Know the dermatologic abnormalities in a patient with cardiac disease and their pathogenesis
  - e. Know the clinical manifestations and significance of embolic phenomena in patients with cardiac disease
  - f. Recognize and understand the importance of variations in hepatic situs
- B. Surface electrocardiography
1. Age-dependent features
    - a. Know age-related changes in ECG wave forms and intervals
  2. Hypertrophy
    - a. Recognize the ECG pattern of ventricular hypertrophy at various ages
    - b. Recognize atrial enlargement on ECG
  3. Depolarization
    - a. Recognize variations from normal pattern of atrial depolarization on ECG and make an interpretation
    - b. Recognize variations from normal pattern of ventricular depolarization on ECG and make an interpretation
    - c. Differentiate the ECG patterns associated with preexcitation, bundle branch block, and hemiblock
  4. Repolarization
    - a. Recognize on ECG variations from normal pattern of ventricular repolarization and make an interpretation
  5. Clinical applications
    - a. Recognize patterns of ischemia, injury, and infarction
  6. Monitoring
    - a. Know the indications for 24-hour ambulatory ECG recording, how to recognize artifacts, and how to interpret the results
    - b. Know the indications for use of an event monitor or an implantable loop recorder and how to interpret the results
- C. Exercise stress testing
1. Know the guidelines for exercise in normal children
  2. Know the guidelines for exercise in children with cardiovascular disease
  3. Recognize the normal responses to exercise in terms of heart rate, blood pressure, cardiac output, oxygen uptake and consumption, and venous return
  4. Understand the ventilatory response to CO<sub>2</sub> in terms of CO<sub>2</sub> response curves and central and peripheral chemoreceptors
  5. Know the indications for and risks of exercise testing in children
  6. Understand the techniques, physiology, advantages, and disadvantages of the different types of exercise (cycle, treadmill, hand-grip exercise)
  7. Understand the physiologic principles related to electrocardiographic responses to exercise
  8. Understand the physiologic principles involved in the ventilatory response to exercise
  9. Understand the indications of nuclear medicine stress testing
- D. Echocardiography/Doppler methods (includes fetal)
1. Assess systolic and diastolic function utilizing echocardiography
  2. Know how to determine gradients and pressure measurement from Doppler-derived velocity measurements

3. Know how Doppler-derived velocity measurements compare to direct-pressure gradient determinations
  4. Understand the principles of echocardiography
  5. Understand the physics of echocardiography
  6. Know the indications for, risks of, and limitations of transesophageal, stress, and fetal echocardiography
  7. Recognize normal cardiac anatomy on echocardiography
- E. Catheterization and intervention
1. General principles
    - a. Know the indications and contraindications for and risks of cardiac catheterization
    - b. Know the most appropriate positional view to obtain optimal angiographic visualization of the targeted cardiac and vasculature structure of interest
    - c. Interpret normal and abnormal pressure data during cardiac catheterization
    - d. Know the normal and potential abnormal courses of a cardiac catheter during cardiac catheterization and angiography
  2. Calculating cardiac output and shunts
    - a. Invasive and noninvasive methods of calculating cardiac output
      1. Know how to calculate myocardial oxygen consumption from data measuring coronary blood flow and oxygen saturation
      2. Understand the concept, use, and limitations of the Fick method to determine blood flow (systemic and pulmonary)
      3. Calculate a right-to-left and a left-to-right shunt given relevant oxygen data
      4. Know how to arrive at a physiologic diagnosis given saturation data
      5. Understand the concept of effective pulmonary blood flow
      6. Recognize important sources of measurement error when quantifying ventricular function by invasive methods
  3. Interventional catheterization: balloon angioplasty/valvuloplasty/stent placement and angiography
    - a. Know which lesions can be dilated by angioplasty
    - b. Understand the factors associated with angioplasty (eg, indications, contraindications, risks, and limitations)
    - c. Know how to perform angioplasty of native and postoperative pulmonary branch stenosis
    - d. Manage the complications of angioplasty
    - e. Interpret patterns of pulmonary and systemic vasculature on angiography
    - f. Understand the factors associated with use of angiography (eg, risks, risk management, complications, and contraindications)
    - g. Know the methods for and limitations of calculations of pulmonary and systemic vascular resistance and its application
    - h. Know which valve lesions are candidates for balloon valvuloplasty
    - i. Understand the factors associated with stent placement (eg, indications, contraindications, risks, and limitations)
    - j. Understand the basic technical aspects of stent placement
    - k. Understand medical management implications following stent placement in lesions
    - l. Identify and manage the complications of stent placement

- m. Understand the factors associated with dilation of bioprosthetic valves/conduits (eg, indications, contraindications, risks, and limitations)
    - n. Understand the factors associated with bioprosthetic valves/conduits (eg, indications, contraindications, risks, and limitations)
    - o. Know how to perform angioplasty of bioprosthetic valves/conduits
  - 4. Transeptal puncture and atrioseptostomy
    - a. Know the indications, contraindications, risks, and limitations of atrioseptostomy
    - b. Understand the basic technical aspects of balloon atrial septostomy
    - c. Understand the factors associated with transeptal puncture (eg, indications, contraindications, risks, and limitations)
  - 5. Occlusion techniques
    - a. General principles
      - 1. Understand the factors associated with occlusion techniques (eg, indications, contraindications, risks, and limitations)
      - 2. Understand the basic technical aspects of occlusion techniques
      - 3. Recognize commonly approved occlusion devices for cardiac defects
      - 4. Plan appropriate management and follow-up evaluation relative to complications of occlusion devices
      - 5. Plan prophylactic management of thrombosis following the use of an occlusion device
  - 6. Percutaneous valve placement
    - a. Understand the principles of percutaneous valve placement
  - 7. Other interventional procedures
    - a. Understand the basic principles and techniques of latest interventional technologies
- F. Cardiac magnetic resonance imaging (MRI) and angiography (MRA) and computed tomography (CT) scan
  - 1. Understand the factors associated with cardiac CT scan and CT angiography (eg, indications, contraindications, and risks)
  - 2. Understand the factors associated with use of cardiac MRI (eg, indications, contraindications, risks, and limitations)
  - 3. Diagnose cardiovascular disease based on cardiac computed tomography
  - 4. Diagnose cardiac disease based on findings on cardiac MRI
  - 5. Know the factors associated with MRI/MRA assessment of cardiovascular function, including flow (eg, indications, techniques, limitations)
- G. Nuclear testing
  - 1. Know the indications, contraindications, risks, and limitations of radionuclide angiocardiology
  - 2. Recognize the clinical implications of normal and abnormal findings on lung perfusion scans and ventilation/perfusion scans
- H. Other forms of cardiac testing
  - 1. Pulse oximetry
    - a. Interpret the principles of pulse oximetry in the evaluation of a patient with cardiovascular disease
    - b. Understand the principles and limitations of pulse oximetry
  - 2. X-ray studies of the chest

- a. Understand the cardiac risks associated with short- and long-term radiation exposure and know how to manage
- I. Electrophysiologic testing
  - 1. General principles
    - a. Measure appropriate intervals during electrophysiologic studies
    - b. Recognize normal and abnormal responses to cardiac pacing as part of electrophysiologic evaluation
    - c. Know indications, contraindications, risks, and limitations for an electrophysiologic study
    - d. Know indications for ablation of supraventricular and ventricular ectopic beats
    - e. Know how to manage the complications of radiofrequency ablation
    - f. Understand the factors associated with radiofrequency ablation (eg, indications, contraindications, risks, and limitations)
    - g. Recognize and understand the mechanism(s) of cardiac arrhythmias induced at the time of electrophysiologic study
    - h. Recognize the types of tachyarrhythmias detected/induced during electrophysiologic studies
    - i. Recognize the site of first-, second-, and third-degree atrioventricular block on an electrophysiologic study
    - j. Recognize the significance of a long H-V interval on an electrophysiologic study
    - k. Recognize electrophysiologic features of atrial and ventricular ectopy
    - l. Understand the effects of different pacing modalities on ventricular function
  - 2. Electrocardiographic diagnosis
    - a. Recognize electrocardiographic features of first-, second-, and third-degree atrioventricular block
    - b. Recognize electrocardiographic features of atrial and ventricular ectopy
- J. Laboratory testing
  - 1. Erythrocyte indices
    - a. Recognize alterations in erythrocyte indices that are important in the evaluation of a patient with cardiovascular disease
  - 2. Arterial blood gases and pH
    - a. Understand factors which influence oxygen-hemoglobin linkage and dissociation curves
    - b. Understand how arterial blood gases and pH are used in the assessment of cardiovascular disease
  - 3. Cardiac injury profiles
    - a. Know the uses of serum creatine kinase activity and troponin I and T concentration measurements in a patient with cardiovascular disease
  - 4. Heart failure profiles
    - a. Understand the use and limitations of biomarkers in the evaluation of acute and chronic heart failure

#### **4. Office-Based Cardiac Problems**

- A. Syncope
  - 1. Know how to evaluate syncope and presyncope
  - 2. Diagnose and manage neurocardiogenic syncope
  - 3. Know the differential diagnosis of syncope

- B. Lipidopathies and coronary risk management
    - 1. Know the various types of hyperlipidemias, including manifestations, their genetic basis, mode of transmission, diagnosis, and management
    - 2. Interpret serum lipid profile tests in children
    - 3. Plan appropriate management of a child with hypercholesterolemia
    - 4. Know the recommended daily dietary saturated fat intake for children of different ages
    - 5. Know the risk factors for hyperlipidemia and the timing of lipid testing based on risk factors
  - C. Hypertension
    - 1. Recognize and plan an appropriate evaluation in a patient with systemic hypertension
    - 2. Plan appropriate management of systemic hypertension
    - 3. Understand the natural history of systemic hypertension in children
  - D. Exercise restrictions in congenital heart disease
    - 1. Know the guidelines for exercise in normal children
    - 2. Know the guidelines for exercise in children with cardiovascular disease
  - E. Chest pain
    - 1. Plan the diagnosis and management of chest pain in children and adolescents
  - F. Cardiovascular problems in the athlete
    - 1. Know how to use echocardiography to recognize physiologic cardiac hypertrophy in an athlete, including differentiation from pathologic hypertrophy
    - 2. Understand and recognize cardiovascular and electrocardiographic changes (eg, heart rate, blood pressure, LV mass) in a well-trained athlete
    - 3. Identify the cause(s) of sudden cardiac death in an athlete
    - 4. Understand utilization and limitations of cardiovascular screening of the athlete and school-aged child
  - G. Cyanosis
    - 1. Recognize manifestations of acute and chronic insufficient pulmonary blood flow
  - H. Palpitations
    - 1. Plan the evaluation and management of palpitations in children and adolescents
  - I. Murmurs (See II.A.3)
5. **Left-to-Right Shunts**
- A. Atrial septal defects (non-AVSD types)
    - 1. Anatomy
      - a. Recognize the anatomic characteristics of different types of atrial septal defects
      - b. Recognize cardiovascular lesions commonly associated with an atrial septal defect
    - 2. Physiology
      - a. Understand the factors that affect shunting at the atrial level
    - 3. Natural history
      - a. Understand the short- and long-term effects of an atrial septal defect on pulmonary vascular bed and cardiac function
      - b. Understand the relationship between an atrial septal defect and arrhythmia
      - c. Understand the relationship between an atrial septal defect and strokes
    - 4. Clinical findings
      - a. Recognize the clinical findings consistent with an atrial septal defect
    - 5. Laboratory findings

- a. Recognize features associated with atrial septal defect using available laboratory tests and recognize important anatomic features that could affect surgical management
  - b. Calculate pulmonary and systemic flows, blood flow ratios, and resistance by various modalities in a patient with atrial septal defect
  - c. Recognize the cardiac MRI features associated with each type of atrial septal defect
- 6. Management, including complications
  - a. Develop an appropriate management plan for a patient with an atrial septal defect
  - b. Determine the appropriate timing of surgical or catheter intervention in a patient with an atrial septal defect
  - c. Identify and manage the early and long-term complications of surgical or catheter closure of an atrial septal defect
- B. Atrioventricular septal defects
  - 1. Embryology, epidemiology, and genetics
    - a. Recognize the genetic syndromes associated with atrioventricular septal defect
  - 2. Anatomy
    - a. Recognize cardiovascular lesions commonly associated with atrioventricular septal defect including varying AVV morphology and insertion
  - 3. Physiology
    - a. Identify the effects of an atrioventricular septal defect on the pulmonary vascular bed
    - b. Understand the factors that determine shunting at atrial and ventricular levels in atrioventricular septal defect
  - 4. Natural history
    - a. Understand the natural history of atrioventricular septal defect
  - 5. Clinical findings
    - a. Recognize the clinical findings consistent with an atrioventricular septal defect
  - 6. Laboratory findings
    - a. Recognize features of atrioventricular septal defects using available laboratory tests and recognize important anatomic features that could affect surgical management
    - b. Evaluate pulmonary pressures in a patient with atrioventricular septal defect
    - c. Recognize the various atrioventricular valve morphologies and attachments in a patient with atrioventricular septal defect
  - 7. Management, including complications
    - a. Recognize implications of straddling and overriding of atrioventricular valves on management decisions
    - b. Plan appropriate medical management of a patient with an atrioventricular septal defect
    - c. Plan the appropriate timing of surgery in a patient with an atrioventricular septal defect
    - d. Recognize and manage early and long-term complications of surgical repair of an atrioventricular septal defect
    - e. Recognize and manage the complications of an unoperated atrioventricular septal defect
- C. Non-AVSD Ventricular septal defects
  - 1. Embryology, epidemiology, and genetics

- a. Recognize specific genetic syndromes associated with ventricular septal defect
- 2. Anatomy
  - a. Know the anatomic location of various types of ventricular septal defects
  - b. Recognize cardiovascular lesions commonly associated with ventricular septal defect
- 3. Physiology
  - a. Identify the effects of a ventricular septal defect on the pulmonary vascular bed
  - b. Understand the effects of ventricular septal defect on cardiac function
  - c. Understand the vascular and cardiac factors that determine shunting in a ventricular septal defect
- 4. Natural history
  - a. Understand the natural history of ventricular septal defect
- 5. Clinical findings
  - a. Differentiate between a closing VSD and development of subpulmonary obstruction
  - b. Recognize and treat pulmonary arterial hypertension
  - c. Recognize the clinical findings associated with a ventricular septal defect
  - d. Recognize the clinical findings of commonly associated cardiovascular lesions with a ventricular septal defect (eg, aortic insufficiency, patent ductus arteriosus, double-chambered right ventricle)
- 6. Laboratory findings
  - a. Recognize the anatomic types of features of ventricular septal defect using available laboratory tests and recognize important anatomic features that could affect surgical management
  - b. Distinguish between restrictive and nonrestrictive communications by Doppler echocardiography in a patient with a ventricular septal defect
- 7. Management, including complications
  - a. Recognize and manage the early and long-term complications following surgical repair or catheter device closure of a ventricular septal defect
  - b. Plan management of a patient with VSD and aortic regurgitation
  - c. Plan appropriate medical management of a patient with ventricular septal defect
  - d. Plan the appropriate timing of surgical or catheter intervention in the management of a patient with ventricular septal defect
  - e. Recognize and manage the complications of unoperated ventricular septal defect
- D. Patent ductus arteriosus
  - 1. Embryology, epidemiology, and genetics
    - a. Know the embryologic basis of patent ductus arteriosus
  - 2. Anatomy
    - a. Recognize the anatomic details of patent ductus arteriosus
    - b. Recognize lesions commonly associated with patent ductus arteriosus
  - 3. Physiology
    - a. Identify the effects of patent ductus arteriosus on the pulmonary vascular bed
    - b. Recognize the effects of gestational age at birth and postnatal age on the presentation of a patent ductus arteriosus
    - c. Understand the effects of a patent ductus arteriosus on cardiac function
    - d. Understand the determinants of shunting in patent ductus arteriosus



4. Natural history
  - a. Understand the natural history of patent ductus arteriosus
5. Clinical findings
  - a. Recognize the clinical findings associated with patent ductus arteriosus
6. Laboratory findings
  - a. Recognize features of patent ductus arteriosus of various sizes using available laboratory tests and recognize important anatomic features that could affect surgical management
  - b. Assess volume overload and estimate pulmonary arterial pressures by echocardiography in a patient with patent ductus arteriosus
7. Management, including complications
  - a. Plan management of patent ductus arteriosus in preterm and term infants, including appropriate use of prostaglandin inhibitors
  - b. Plan the timing of surgical or catheter intervention in a patient with patent ductus arteriosus
  - c. Recognize possible early and long-term complications of surgical or transcatheter repair of patent ductus arteriosus
  - d. Recognize and manage the complications of untreated patent ductus arteriosus
  - e. Understand the relationship between patent ductus arteriosus and necrotizing enterocolitis in preterm and full-term infants
- E. Coronary arteriovenous fistula
  1. Anatomy
    - a. Understand the pathologic features of a coronary arteriovenous fistula
  2. Physiology
    - a. Understand the vascular physiology of a large coronary arteriovenous fistula
  3. Natural history
    - a. Understand the natural history of coronary arteriovenous fistula
  4. Clinical findings
    - a. Recognize the clinical findings of coronary arteriovenous fistula
  5. Laboratory findings
    - a. Recognize features associated with coronary arteriovenous fistula using available laboratory tests and recognize important anatomic features that could affect surgical management
  6. Management, including complications
    - a. Plan the appropriate technique and timing of surgical or catheter intervention in a patient with coronary arteriovenous fistula
    - b. Identify and manage possible complications of surgical or transcatheter repair of coronary arteriovenous fistula
    - c. Recognize and manage the complications of an untreated arteriovenous fistula
- F. Aortopulmonary window
  1. Embryology, epidemiology, and genetics
    - a. Know the embryologic basis of aortopulmonary window
  2. Anatomy
    - a. Recognize the anatomic details of an aortopulmonary window
    - b. Recognize lesions commonly associated with an aortopulmonary window
  3. Physiology

- a. Identify the effects of an aortopulmonary window on the pulmonary vascular bed
- 4. Natural history
  - a. Understand the natural history of an unoperated aortopulmonary window
- 5. Clinical findings
  - a. Recognize the clinical presentation of aortopulmonary window
- 6. Laboratory findings
  - a. Recognize features consistent with a diagnosis of aortopulmonary window by available laboratory tests and recognize important features that could affect surgical management
- 7. Management, including complications
  - a. Plan the appropriate timing of surgical or catheter intervention in a patient with aortopulmonary window
  - b. Recognize and manage the long-term complications of unoperated aortopulmonary window
- 6. **Right-to-Left Shunts**
  - A. Tetralogy of Fallot
    - 1. Embryology, epidemiology, and genetics
      - a. Know the embryology, epidemiology, and genetics for tetralogy of Fallot
      - b. Recognize genetic syndromes associated with tetralogy of Fallot
    - 2. Anatomy
      - a. Recognize the commonly associated lesions in a patient with tetralogy of Fallot
      - b. Recognize the anatomic features in a patient with tetralogy of Fallot
    - 3. Physiology
      - a. Understand the circulatory abnormalities in a patient with tetralogy of Fallot, including the pathophysiology of a hypercyanotic episode
    - 4. Natural history
      - a. Know the natural history of tetralogy of Fallot
    - 5. Clinical findings
      - a. Recognize the clinical features of hypercyanotic episodes in patients with tetralogy of Fallot
    - 6. Laboratory findings
      - a. Appropriately use and interpret diagnostic studies for evaluation and treatment planning in tetralogy of Fallot
      - b. Recognize hemodynamic and angiographic findings of tetralogy of Fallot
    - 7. Management, including complications
      - a. Plan management of a hypercyanotic episode in a patient with tetralogy of Fallot
      - b. Plan the treatment approach for palliation or correction of tetralogy of Fallot
      - c. Recognize the complications of cyanosis in untreated tetralogy of Fallot
      - d. Recognize and manage complications after treatment of tetralogy of Fallot
  - B. Double-outlet right ventricle
    - 1. Embryology, epidemiology, and genetics
      - a. Understand the etiology, epidemiology, and genetic syndromes associated with double-outlet right ventricle
    - 2. Anatomy
      - a. Recognize the anatomic features of double-outlet right ventricle and commonly associated lesions

3. Physiology
    - a. Understand the circulatory physiology of double-outlet right ventricle and its relationship with anatomic features
  4. Laboratory findings
    - a. Appropriately use and interpret diagnostic studies for evaluation and treatment planning in double-outlet right ventricle
  5. Management, including complications
    - a. Recognize the effects of the transitional circulation on the clinical presentation of double-outlet right ventricle
    - b. Plan the treatment of double-outlet right ventricle based on anatomic and physiological variables
    - c. Recognize and manage the early and long-term complications after therapy in a patient with double-outlet right ventricle
- C. Pulmonary atresia with intact septum
1. Embryology, epidemiology, and genetics
    - a. Know the embryology, epidemiology, and genetics of pulmonary atresia with intact septum
    - b. Recognize the etiology, epidemiology, and genetic syndromes associated with pulmonary atresia with intact septum
  2. Anatomy
    - a. Recognize the anatomic features and their prognostic significance in pulmonary atresia with intact septum
    - b. Recognize cardiovascular lesions commonly associated with pulmonary atresia with intact septum
  3. Physiology
    - a. Understand the physiologic consequences of the anatomic spectrum of pulmonary atresia with intact septum
  4. Natural history
    - a. Understand the range of natural history in pulmonary atresia with intact septum
  5. Laboratory findings
    - a. Appropriately use and interpret diagnostic studies for evaluation and treatment planning in pulmonary atresia with intact ventricular septum before and after intervention.
  6. Management, including complications
    - a. Plan the treatment of pulmonary atresia with intact septum
    - b. Recognize and manage early and long-term complications of therapy in pulmonary atresia with intact septum
- D. Pulmonary atresia with ventricular septal defect
1. Embryology, epidemiology, and genetics
    - a. Know the embryology, epidemiology, and genetics of pulmonary atresia with ventricular septal defect
    - b. Recognize the etiology, epidemiology, and genetic syndromes associated with pulmonary atresia with ventricular septal defect
  2. Anatomy
    - a. Recognize the abnormalities of the pulmonary vascular bed in pulmonary atresia with ventricular septal defect

- b. Recognize lesions commonly associated with pulmonary atresia with ventricular septal defect
  - 3. Physiology
    - a. Understand the consequences of anatomic variants of pulmonary atresia with ventricular septal defect on treatment and prognosis
  - 4. Natural history
    - a. Understand the range of natural history in pulmonary atresia with ventricular septal defect
  - 5. Laboratory findings
    - a. Recognize features associated with pulmonary atresia with ventricular septal defect using available laboratory tests and recognize important anatomic features that could affect surgical management
  - 6. Management, including complications
    - a. Plan the treatment of pulmonary atresia with ventricular septal defect
    - b. Recognize and manage early and long-term complications of therapy in pulmonary atresia with ventricular septal defect
    - c. Recognize and manage a patient with unoperated pulmonary atresia and ventricular septal defect
- E. Absent pulmonary valve syndrome
  - 1. Embryology, epidemiology, and genetics
    - a. Recognize the etiology, epidemiology, and genetic syndromes associated with absent pulmonary valve syndrome
  - 2. Anatomy
    - a. Recognize the anatomic features and associated lesions in a patient with absent pulmonary valve syndrome
  - 3. Physiology
    - a. Understand the physiologic abnormalities in absent pulmonary valve syndrome
  - 4. Natural history
    - a. Understand the natural history of absent pulmonary valve syndrome
  - 5. Clinical findings
    - a. Recognize the clinical features of absent pulmonary valve syndrome
  - 6. Laboratory findings
    - a. Appropriately use and interpret diagnostic studies for evaluation and treatment planning in pulmonary atresia with absent pulmonary valve syndrome before and after intervention.
  - 7. Management, including complications
    - a. Plan the surgical approach for palliation or correction of absent pulmonary valve syndrome
    - b. Recognize and manage the short- and long-term airway complications associated with absent pulmonary valve syndrome
- 7. **Single Ventricular Lesions**
  - A. Hypoplastic left heart syndrome
    - 1. Embryology, epidemiology, and genetics
      - a. Recognize the etiology, epidemiology, and genetic syndromes associated with hypoplastic left heart syndrome
    - 2. Anatomy

- a. Identify anatomic features of hypoplastic left heart syndrome
- 3. Physiology
  - a. Understand the physiology of hypoplastic left heart syndrome
  - b. Understand the circulatory and metabolic effects of hypoplastic left heart syndrome
  - c. Understand the impact of hypoplastic left heart syndrome and its variants on fetal pulmonary and neurodevelopment.
- 4. Clinical findings
  - a. Recognize the range of clinical presentation of hypoplastic left heart syndrome and its evolution after birth
  - b. Recognize features of hypoplastic left heart syndrome using available laboratory tests and recognize important anatomic features that could affect surgical management
- 5. Management, including complications
  - a. Plan appropriate management of obstructed atrial septum in a patient with hypoplastic left heart syndrome
  - b. Recognize the treatment options for hypoplastic left heart syndrome, their merits and drawbacks.
  - c. Plan the management of a neonate with hypoplastic left heart syndrome who has undergone Norwood palliation
  - d. Plan the management of an infant with hypoplastic left heart syndrome who has undergone hybrid palliation
- B. Tricuspid atresia
  - 1. Etiology, epidemiology, and genetics
    - a. Understand the embryologic basis of tricuspid atresia
  - 2. Anatomy
    - a. Identify anatomic features of tricuspid atresia and associated lesions
  - 3. Physiology
    - a. Understand the circulatory physiology of tricuspid atresia
  - 4. Natural history
    - a. Understand the natural history of tricuspid atresia
  - 5. Clinical findings
    - a. Recognize the clinical findings in a patient with tricuspid atresia
  - 6. Laboratory findings
    - a. Recognize features of tricuspid atresia using available laboratory tests and recognize important anatomic features that could affect surgical management
  - 7. Management, including complications
    - a. Plan appropriate medical management for a patient with tricuspid atresia
    - b. Diagnose and manage the postoperative complications following surgical palliation of tricuspid atresia
- C. Other forms of single (univentricular) heart
  - 1. Etiology, epidemiology, and genetics
    - a. Understand the developmental basis for a univentricular heart
  - 2. Anatomy
    - a. Identify anatomic features of the various forms of univentricular heart
  - 3. Physiology
    - a. Understand the physiology of the univentricular heart

4. Natural history
  - a. Understand the natural history of univentricular hearts and the impact on surgical decisions
5. Clinical findings
  - a. Recognize the typical clinical findings of univentricular hearts
6. Laboratory findings
  - a. Recognize features associated with univentricular heart using available laboratory tests and recognize important anatomic features that could affect surgical management
7. Management, including complications
  - a. Plan medical management of a patient with univentricular heart
  - b. Plan surgical management of a patient with a univentricular heart
  - c. Manage complications of the natural history and medical therapy of a univentricular heart
  - d. Manage complications of neonatal surgical palliations in univentricular hearts after treatment
- D. The Glenn and Fontan Circulation
  1. Etiology, epidemiology, and genetics
    - a. Recognize the indications and contraindications to Glenn and Fontan operations
  2. Anatomy
    - a. Identify anatomical variants of Glenn and Fontan circulation
  3. Physiology
    - a. Know the physiologic effects of Glenn and Fontan circulation on the heart and circulation
  4. Clinical findings
    - a. Recognize the typical clinical features of a Glenn and Fontan circulation
  5. Laboratory findings
    - a. Recognize features associated with Glenn and Fontan circulation using available laboratory tests and recognize important anatomic features that could affect management
  6. Management, including complications
    - a. Recognize and manage the early and long-term complications of a patient with Glenn and Fontan circulation, such as protein-losing enteropathy
8. **Structural, Valvar, and Obstructive Lesions**
  - A. Tricuspid valve
    1. Tricuspid stenosis and regurgitation
      - a. Etiology, epidemiology, and genetics
        1. Recognize the etiology of tricuspid regurgitation
      - b. Anatomy
        1. Recognize lesions commonly associated with tricuspid stenosis and regurgitation
      - c. Physiology
        1. Know the physiologic effects of tricuspid stenosis and regurgitation on the heart and circulation in a neonate
      - d. Natural history
        1. Understand the natural history of tricuspid stenosis and regurgitation

- e. Clinical findings
  - 1. Recognize typical clinical features in a patient with tricuspid stenosis and/or regurgitation
- f. Laboratory findings
  - 1. Recognize and interpret features of tricuspid valve stenosis and regurgitation using available laboratory tests and recognize important anatomic features that could affect surgical management
- g. Management, including complications
  - 1. Plan appropriate medical and surgical therapy in a patient with tricuspid stenosis and regurgitation
- 2. Ebstein anomaly of the tricuspid valve
  - a. Etiology, epidemiology, and genetics
    - 1. Know the embryologic basis for Ebstein anomaly of the tricuspid valve
  - b. Anatomy
    - 1. Recognize pathologic features of Ebstein anomaly of the tricuspid valve and associated lesions
  - c. Physiology
    - 1. Know the spectrum of abnormalities in Ebstein anomaly of the tricuspid valve
  - d. Natural history
    - 1. Know the natural history of a patient with Ebstein anomaly of the tricuspid valve
  - e. Clinical findings
    - 1. Recognize Ebstein anomaly of the tricuspid valve based on clinical findings
  - f. Laboratory findings
    - 1. Recognize features of Ebstein anomaly of the tricuspid valve using available laboratory tests and recognize important anatomic features that could affect surgical management
  - g. Management, including complications
    - 1. Understand physiology of and plan medical management of a neonate with Ebstein anomaly of the tricuspid valve and severe hypoxemia
    - 2. Plan appropriate interventional therapy in a patient with Ebstein anomaly of the tricuspid valve
- B. Obstructive lesions of the right ventricular outflow tract, pulmonary valve, and pulmonary branches
  - 1. Double-chamber right ventricle
    - a. Anatomy
      - 1. Identify anatomical features of double-chamber right ventricle
      - 2. Recognize lesions commonly associated with double-chamber right ventricle
    - b. Physiology
      - 1. Know the physiologic effects of double-chamber right ventricle on the heart and circulation in a neonate
      - 2. Understand the physiology of double-chamber right ventricle in a neonate
    - c. Natural history
      - 1. Know the natural history of a patient with double-chamber right ventricle
    - d. Clinical findings

1. Recognize the typical clinical features of double-chamber right ventricle in a neonate and older child
  - e. Laboratory findings
    1. Recognize features associated with double-chamber right ventricle using available laboratory tests and recognize important anatomic features that could affect surgical management
  - f. Management, including complications
    1. Plan management of a patient with double-chamber right ventricle
    2. Recognize and manage the early and long-term complications of surgical intervention in a patient with double-chamber right ventricle
2. Pulmonary valve stenosis
  - a. Embryology, epidemiology, and genetics
    1. Recognize the embryology, epidemiology, and genetics associated with pulmonary valve stenosis
  - b. Anatomy
    1. Identify abnormalities of pulmonary valve structure
    2. Recognize lesions commonly associated with pulmonary valve stenosis
  - c. Physiology
    1. Know the physiologic effects of pulmonary valve stenosis on the heart and circulation in a neonate
    2. Understand the physiology of critical pulmonary valve stenosis in a neonate
  - d. Natural history
    1. Know the natural history of a patient with pulmonary valve stenosis
  - e. Clinical findings
    1. Recognize the typical clinical features of pulmonary valve stenosis in a neonate and older child
  - f. Laboratory findings
    1. Recognize features associated with pulmonary valve stenosis using available laboratory tests and recognize important anatomic features that could affect surgical management
  - g. Management, including complications
    1. Plan management of a patient with pulmonary valve stenosis
    2. Recognize and manage the early and long-term complications of surgical and transcatheter intervention in a patient with pulmonary valve stenosis
3. Pulmonary regurgitation
  - a. Embryology, epidemiology, and genetics
    1. Recognize the etiology, epidemiology, and genetic syndromes associated with pulmonary regurgitation
    2. Recognize lesions commonly associated with pulmonary regurgitation
    3. Know the effects of pulmonary regurgitation on cardiovascular physiology
    4. Recognize the natural history of untreated chronic pulmonary regurgitation
  - b. Clinical findings
    1. Recognize the typical clinical findings of pulmonary regurgitation
    2. Recognize the indications for pulmonary valve replacement in pulmonary regurgitation
  - c. Management, including complications



1. Recognize and manage complications of pulmonary valve replacement
4. Pulmonary branch stenosis
  - a. Embryology, epidemiology, and genetics
    1. Understand the embryologic basis of the formation of the proximal and distal pulmonary vascular bed
    2. Recognize the genetic syndromes associated with pulmonary artery stenosis
  - b. Anatomy
    1. Recognize cardiac lesions commonly associated with pulmonary artery stenosis
  - c. Physiology
    1. Know the physiologic effects of pulmonary branch stenosis on the heart and circulation in a neonate
  - d. Natural history
    1. Understand the effect of genetic syndrome on the natural history of pulmonary artery stenosis
  - e. Clinical findings
    1. Recognize the clinical findings of pulmonary artery stenosis
  - f. Laboratory findings
    1. Recognize features of pulmonary stenosis using available diagnostic tests and recognize important anatomic features that could affect surgical management
  - g. Management, including complications
    1. Plan appropriate management of a patient with pulmonary artery stenosis
    2. Recognize and treat the complications of therapy for pulmonary artery stenosis
- C. Mitral stenosis, mitral regurgitation, and mitral valve prolapse
  1. Mitral stenosis
    - a. Anatomy
      1. Recognize anatomic features of mitral stenosis
    - b. Physiology
      1. Recognize the physiology of anomalies of the mitral valve on the pulmonary and systemic circulations
    - c. Natural history
      1. Understand the natural history of mitral stenosis
    - d. Clinical findings
      1. Recognize the clinical features of the various forms of mitral stenosis
    - e. Laboratory findings
      1. Recognize mitral stenosis using available laboratory tests and recognize important anatomic features that could affect surgical management
    - f. Management, including complications
      1. Recognize the complications that may occur in association with medical, transcatheter, or surgical management of mitral stenosis
  2. Mitral regurgitation
    - a. Etiology, epidemiology, and genetics
      1. Know the etiology, epidemiology, and genetics of mitral regurgitation
    - b. Anatomy
      1. Recognize anatomic features of mitral regurgitation
    - c. Physiology

1. Recognize the effect of mitral regurgitation on pulmonary vascular resistance and systemic circulations
- d. Natural history
  1. Understand the natural history of mitral valve regurgitation
- e. Clinical findings
  1. Recognize the clinical features in a patient with mitral regurgitation
- f. Laboratory findings
  1. Recognize features of mitral regurgitation using available laboratory tests and recognize important anatomic features that could affect surgical management
- g. Management, including complications
  1. Recognize and manage the complications that may occur following medical or surgical management of mitral regurgitation
3. Mitral valve prolapse
  - a. Etiology, epidemiology, and genetics
    1. Know the etiology, epidemiology, and genetics of mitral valve prolapse
    2. Know mode of genetic transmission of mitral valve prolapse
  - b. Anatomy
    1. Identify anatomic features of mitral valve prolapse
    2. Recognize lesions commonly associated with mitral valve prolapse
  - c. Physiology
    1. Know the physiologic effects of mitral valve prolapse on the heart and circulation in a neonate
    2. Understand the physiology of critical mitral valve prolapse in a neonate
  - d. Natural history
    1. Know the natural history of a patient with mitral valve prolapse
  - e. Clinical findings
    1. Recognize the typical clinical features of mitral valve prolapse in a neonate and older child
    2. Know the provocative maneuvers that increase mitral regurgitation in a patient with mitral valve prolapse
  - f. Laboratory findings
    1. Recognize features associated with mitral valve prolapse using available laboratory tests and recognize important anatomic features that could affect surgical management
  - g. Management, including complications
    1. Plan management of a patient with mitral valve prolapse
    2. Recognize and manage the early and long-term complications of surgical intervention in a patient with mitral valve prolapse
    3. Recognize the echocardiographic features of mitral valve prolapse using available diagnostic tests and recognize important anatomic features that could affect surgical management
- D. Obstructive lesions of the left ventricular outflow tract, aortic valve, and aortic arch
  1. Common embryology, epidemiology and genetics of LVOT, aortic valve, and aortic arch lesions
    - a. Know the embryology of subaortic stenosis
    - b. Know the embryology of aortic valve stenosis and bicuspid aortic valve

- c. Know the embryology of supravalvar aortic stenosis
  - d. Know the embryology of aortic coarctation, including the association between neural crest development and interrupted aortic arch
  - e. Recognize the epidemiology and genetics of the abnormalities of aortic arches (including anomalous systemic arteries)
  - f. Know the epidemiology of left ventricular outflow tract lesions
  - g. Know the genetic syndromes associated with left ventricular outflow tract lesions
  - h. Recognize the spectrum of presentation of left ventricular outflow tract obstruction based on age and severity
2. Subaortic stenosis
- a. Anatomy
    - 1. Recognize the anatomic types and features of various forms of subaortic stenosis
    - 2. Recognize the lesions commonly associated with subaortic stenosis
  - b. Natural history
    - 1. Understand the natural history of subaortic stenosis
  - c. Physiology
    - 1. Know the effects of changing physiology on degree of subaortic stenosis
  - d. Laboratory findings
    - 1. Recognize laboratory tests used to assess subaortic stenosis
  - e. Management, including complications
    - 1. Plan medical management of a patient who has subaortic stenosis across all age groups
    - 2. Know the indications for medical, transcatheter and surgical intervention of the various types of subaortic stenosis
    - 3. Recognize the early and long-term surgical complications in a patient with subaortic stenosis
3. Bicuspid aortic valve
- a. Anatomy
    - 1. Identify anatomic abnormalities of bicuspid aortic valve
    - 2. Recognize lesions commonly associated with bicuspid aortic valve
  - b. Physiology
    - 1. Know the physiologic effects of bicuspid aortic valve on the heart and circulation in a neonate
    - 2. Understand the physiology of critical bicuspid aortic valve in a neonate
  - c. Natural history
    - 1. Know the natural history of a patient with bicuspid aortic valve
  - d. Clinical findings
    - 1. Recognize the typical clinical features of bicuspid aortic valve in a neonate and older child
  - e. Laboratory findings
    - 1. Recognize features associated with a bicuspid aortic valve using available laboratory tests and recognize important anatomic features that could affect surgical management
  - f. Management, including complications
    - 1. Plan management of a patient with a bicuspid aortic valve

2. Recognize and manage the early and long-term complications of surgical and transcatheter intervention in a patient with bicuspid aortic valve
4. Aortic valve stenosis and regurgitation
  - a. Anatomy
    1. Recognize the anatomic types of aortic valve stenosis including critical aortic stenosis
    2. Recognize the lesions commonly associated with aortic stenosis/regurgitation
  - b. Natural history
    1. Understand the natural history of aortic stenosis/regurgitation including that of critical aortic stenosis and progression of disease
  - c. Physiology
    1. Know the circulatory, metabolic, and myocardial effects of aortic stenosis/regurgitation
  - d. Laboratory findings
    1. Recognize aortic stenosis and regurgitation using available laboratory tests and recognize important anatomic features that could affect surgical management
  - e. Management, including complications
    1. Plan medical management of a patient who has aortic stenosis/insufficiency across various age groups including medical, interventional and surgical approaches
    2. Know the risks and benefits of available interventional and surgical strategies for the treatment of aortic regurgitation based on patient characteristics
    3. Recognize the early and long-term surgical complications in a patient with aortic stenosis/regurgitation, including the risks associated with prosthetic valves
5. Supravalvar aortic stenosis
  - a. Anatomy
    1. Identify anatomical abnormalities of supravalvar aortic stenosis
    2. Recognize lesions commonly associated with supravalvar aortic stenosis including coronary artery, pulmonary arterial and renal abnormalities
  - b. Physiology
    1. Understand the circulatory, metabolic, and myocardial effects of supravalvar aortic stenosis including effects on coronary flow
    2. Know the physiologic effects of supravalvar aortic stenosis on the heart and circulation in a neonate
    3. Understand the physiology of critical supravalvar aortic stenosis in a neonate
  - c. Natural history
    1. Know the natural history of a patient with supravalvar aortic stenosis
    2. Recognize the typical clinical features of supravalvar aortic stenosis in a neonate and older child
  - d. Laboratory findings
    1. Recognize features associated with supravalvar aortic stenosis using available laboratory tests and recognize important anatomic features that could affect surgical management
  - e. Management, including complications
    1. Plan management of a patient with supravalvar aortic stenosis

2. Recognize and manage the early and long-term complications of surgical and transcatheter intervention in a patient with supravalvar aortic stenosis
6. Coarctation of the aorta, including interrupted aortic arch and other forms of arch obstruction
  - a. Anatomy
    1. Recognize the anatomic details of coarctation of the aorta and frequently associated lesions
    2. Recognize the anatomic details of interruption of the aortic arch
    3. Recognize commonly associated lesions with interruption of the aortic arch
  - b. Physiology
    1. Identify the circulatory, metabolic, and myocardial abnormalities in a neonate and older child with coarctation of the aorta
  - c. Natural history
    1. Understand the natural history of coarctation of the aorta
  - d. Clinical findings
    1. Recognize the clinical features of interruption of the aortic arch
    2. Recognize the clinical findings in coarctation of the aorta in patients of varying ages
  - e. Laboratory findings
    1. Recognize features of interruption of aortic arch using available laboratory tests and recognize important anatomic features that could affect surgical management.
    2. Recognize features consistent with coarctation of the aorta in the infant and older child using available laboratory tests and recognize important anatomic features that could affect surgical management
    3. Recognize the limitations of echocardiography in the diagnosis of coarctation of the aorta
  - f. Management, including complications
    1. Plan medical and surgical management of interruption of the aortic arch
    2. Recognize the immediate and long-term complications associated with interruption of the aortic arch
    3. Plan the surgical and transcatheter management of a patient with coarctation of the aorta
    4. Recognize and manage the early and long-term complications of surgical and transcatheter repair of coarctation of the aorta
    5. Know the appropriate medical management of patients with treated coarctation of the aorta

## 9. **Congenital Abnormalities of the Great Arteries and Aorta**

### A. Truncus arteriosus

1. Embryology, epidemiology, and genetics
  - a. Understand the embryologic basis of truncus arteriosus
  - b. Understand the etiology, epidemiology, and genetic implications of truncus arteriosus
2. Anatomy
  - a. Recognize the features and anatomic details of truncus arteriosus and commonly associated lesions

3. Physiology
    - a. Understand the circulatory physiology in truncus arteriosus
  4. Natural history
    - a. Understand the natural history of a patient with truncus arteriosus
  5. Clinical findings
    - a. Recognize the typical clinical findings of truncus arteriosus
  6. Laboratory findings
    - a. Recognize truncus arteriosus using available laboratory tests and recognize important anatomic features that could affect surgical management
  7. Management, including complications
    - a. Plan the surgical approach for palliation or correction of truncus arteriosus
    - b. Plan appropriate preoperative medical management of a patient with truncus arteriosus
    - c. Recognize and manage early and long-term postoperative complications following repair of truncus arteriosus
- B. Transposition of the great arteries
1. Embryology, epidemiology, and genetics
    - a. Recognize the etiology, epidemiology, and genetic syndromes associated with transposition of the great arteries
  2. Anatomy
    - a. Recognize the anatomic features and commonly associated lesions in transposition of the great arteries
  3. Physiology
    - a. Understand the cardiac physiology of transposition of the great arteries
  4. Natural history
    - a. Understand the natural history of transposition of the great arteries
  5. Clinical findings
    - a. Recognize the typical clinical findings of transposition of the great arteries
    - b. Recognize the typical presentation of transposition of the great arteries
  6. Laboratory findings
    - a. Recognize features of transposition of the great arteries using available laboratory tests and recognize important anatomic features that could affect surgical management
  7. Management, including complications
    - a. Recognize coronary obstruction following arterial repair of d-transposition of the great arteries
    - b. Recognize and manage the immediate and long-term complications of a severely hypoxemic patient with transposition of the great arteries
    - c. Plan the appropriate surgical management for transposition of the great arteries based on patient characteristics
- C. Congenitally corrected transposition of the great arteries
1. Embryology, epidemiology, and genetics
    - a. Understand the etiology, epidemiology, and genetic syndromes associated with the l-bulboventricular loop and its sequelae
  2. Anatomy

- a. Recognize the anatomic features in l-transposition of the great arteries and commonly associated lesions
- 3. Physiology
  - a. Understand the cardiac physiology of l-transposition of the great arteries
- 4. Natural history
  - a. Understand the natural history of l-transposition of the great arteries
- 5. Clinical findings
  - a. Recognize the typical clinical findings of l-transposition of the great arteries
- 6. Laboratory findings
  - a. Recognize features of l-transposition of the great arteries using available laboratory tests and recognize important anatomic features that could affect surgical management
- 7. Management, including complications
  - a. Plan the surgical approach for palliation or correction of l-transposition of the great arteries
  - b. Recognize and manage the early and long-term postoperative complications of l-transposition of the great arteries
- D. Aortic root and arch abnormalities
  - 1. Management, including complications
    - a. Recognize and manage root and arch abnormalities associated with connective tissue abnormalities
  - 2. Vascular rings and slings
    - a. Embryology, epidemiology, and genetics
      - 1. Understand the etiology, epidemiology, and genetic syndromes associated with vascular rings
      - 2. Understand the embryology of pulmonary artery slings
    - b. Anatomy
      - 1. Know the various types of vascular rings and slings, and their association with airway pathology
      - 2. Know the anatomic details of left pulmonary artery sling and vascular rings
    - c. Physiology
      - 1. Understand the physiology of vascular rings and slings
    - d. Natural history
      - 1. Understand the natural history of vascular rings and slings
    - e. Clinical findings
      - 1. Recognize the clinical presentation of vascular rings and anomalous left pulmonary artery (PA sling), and plan appropriate diagnostic evaluation
    - f. Laboratory findings
      - 1. Recognize features of vascular ring/anomalous left pulmonary artery (PA sling) using available laboratory tests and recognize important anatomic features that could affect surgical management
    - g. Management, including complications
      - 1. Know the indications for surgery for vascular rings
      - 2. Recognize the complications related to repair of vascular rings and slings, including persistent airway problems
- E. Congenital malformations of the coronary circulation

1. Anomalous left coronary artery from the pulmonary artery
  - a. Etiology, epidemiology, and genetics
    1. Know the etiology, epidemiology, and genetics of the coronary artery origin abnormalities
  - b. Anatomy
    1. Know the gross and histologic features and natural history of anomalous left coronary artery and myocardial infarction
  - c. Physiology
    1. Recognize the pathophysiologic effects of anomalous origin of the left coronary artery from the pulmonary artery
  - d. Natural history
    1. Know the natural history of anomalous origin of left coronary artery from the pulmonary trunk, as well as other well-characterized coronary artery anomalies
  - e. Clinical findings
    1. Recognize the various clinical presentations of anomalous origin of left coronary artery from the pulmonary artery
  - f. Laboratory findings
    1. Recognize features of anomalous origin of the left coronary artery from the pulmonary artery using available laboratory tests and recognize important anatomic features that could affect surgical management
  - g. Management
    1. Plan management for anomalous left coronary artery from the pulmonary artery
    2. Understand and manage complications following anomalous left coronary artery from the pulmonary artery
2. Other congenital malformations of the coronary system
  - a. Etiology, epidemiology, and genetics
    1. Know the etiology, epidemiology, and genetics of various coronary artery anomalies (eg, coronary fistula, single coronary artery, coronary artery arising from the wrong sinus, etc)
  - b. Anatomy
    1. Know the gross and histologic features and natural history of various coronary artery anomalies
  - c. Natural history
    1. Know the natural history of various well-characterized coronary artery anomalies
  - d. Clinical findings
    1. Recognize the various clinical presentations of various coronary artery anomalies
  - e. Laboratory findings
    1. Recognize features of various coronary artery anomalies using available laboratory tests and recognize important anatomic features that could affect surgical management
  - f. Management
    1. Understand and manage the clinical course following medical or surgical management for congenital coronary arteries

## **10. Systemic and Pulmonary Venous Abnormalities and Situs Abnormalities**



- A. Systemic venous abnormalities
  - 1. Anatomy
    - a. Be able to identify abnormalities of the systemic veins
    - b. Recognize lesions commonly associated with vena cava abnormalities
  - 2. Physiology
    - a. Know the physiologic effects of systemic venous abnormalities
    - b. Understand the physiologic effects that can lead to acquired systemic venous abnormalities
  - 3. Natural history
    - a. Know the natural history of a patient with vena cava abnormalities
  - 4. Clinical findings
    - a. Recognize the typical clinical features of vena cava abnormalities in a neonate and older child
  - 5. Laboratory findings
    - a. Recognize systemic venous abnormalities (eg, persistent left superior vena cava) using available laboratory tests and recognize important anatomic features that could affect management
  - 6. Management, including complications
    - a. Plan the management of a patient with systemic venous abnormalities
- B. Abnormalities of the pulmonary venous system
  - 1. Anomalous pulmonary venous connection
    - a. Etiology, epidemiology, and genetics
      - 1. Know the embryologic sequence for normal pulmonary venous development
      - 2. Know the embryologic mechanisms responsible for the various forms of anomalous pulmonary venous connections
  - 2. Anatomy
    - a. Identify the anatomy of the various types of partial and total anomalous pulmonary venous connections and associated defects
    - b. Know the anatomical lesions commonly associated with anomalous pulmonary venous connection
  - 3. Physiology
    - a. Understand the physiology of the various types of obstructed and non-obstructed total anomalous pulmonary venous connections
    - b. Understand the circulatory physiology in patients with partial anomalous pulmonary venous connections
  - 4. Natural history
    - a. Understand the natural history of partial anomalous pulmonary venous connections
    - b. Understand the natural history of unobstructed and unobstructed total anomalous pulmonary venous connections
  - 5. Clinical findings
    - a. Recognize the clinical findings of partial anomalous pulmonary venous connections
    - b. Recognize the clinical features of total anomalous pulmonary venous connection, including differences between patients with obstruction and those without obstruction
  - 6. Laboratory findings

- a. Be able to diagnose partial anomalous pulmonary venous connections using available laboratory tests and recognize important anatomic features that could affect surgical management
  - b. Be able to diagnose non-obstructed and obstructed total anomalous pulmonary venous connections using available laboratory tests and recognize important anatomic features that could affect surgical management
- 7. Management, including complications
  - a. Plan the management of a patient with a partial anomalous pulmonary venous connection
  - b. Plan surgical management of a patient with total anomalous pulmonary venous connection based on anatomic features
  - c. Recognize and manage early and long-term complications of surgical repair of total anomalous pulmonary venous connection
  - d. Recognize and manage short and long-term complications following repair of anomalous pulmonary venous connections
- C. Cor triatriatum
  - 1. Etiology, epidemiology, and genetics
    - a. Know the embryologic basis of cor triatriatum
  - 2. Anatomy
    - a. Recognize the various forms of cor triatriatum and the anatomical relationship to left atrial anatomy
  - 3. Physiology
    - a. Understand the physiology of cor triatriatum
  - 4. Natural history
    - a. Understand the natural history of cor triatriatum
  - 5. Clinical findings
    - a. Recognize the clinical presentation of cor triatriatum
  - 6. Laboratory findings
    - a. Diagnose cor triatriatum using available laboratory tests and recognize important anatomic features that could affect surgical management
  - 7. Management, including complications
    - a. Plan medical and surgical management of a patient with cor triatriatum
- D. Pulmonary venous stenosis/atresia
  - 1. Etiology, epidemiology, and genetics
    - a. Know the embryology of pulmonary vein stenosis/atresia
  - 2. Anatomy
    - a. Recognize the anatomic features of pulmonary venous stenosis/atresia and associated lesions
  - 3. Physiology
    - a. Recognize the implications of increasing pulmonary blood flow in a patient with occult pulmonary venous obstruction
    - b. Understand the circulatory effects of pulmonary venous stenosis/atresia
  - 4. Natural history
    - a. Understand the natural history of pulmonary venous stenosis/atresia
  - 5. Clinical findings
    - a. Recognize the clinical features of pulmonary venous stenosis/atresia

6. Laboratory findings
    - a. Recognize and interpret hemodynamic and angiographic findings in a patient with pulmonary venous stenosis/atresia using available laboratory tests and recognize important anatomic features that could affect surgical management
  7. Management, including complications
    - a. Plan the medical and transcatheter or surgical management for a patient with pulmonary venous stenosis/atresia
    - b. Understand complications that may occur with therapy of pulmonary venous stenosis/atresia
- E. Situs abnormalities and heterotaxy syndromes
1. Etiology, epidemiology, and genetics
    - a. Understand the etiology, epidemiology, and embryology of situs abnormalities and relationships between cardiac and visceral situs
  2. Anatomy
    - a. Know anatomic features and variations in atrial situs and commonly associated lesions
    - b. Recognize the anatomic features of superior-inferior ventricles and criss-cross hearts
  3. Physiology
    - a. Understand physiologic consequences of lesions associated with asplenia/polysplenia syndromes
  4. Natural history
    - a. Recognize features of abnormal atrial and visceral situs using available diagnostic tests and recognize important anatomic features that could affect surgical management
    - b. Know the prognosis and natural history of patients with the major forms of situs abnormalities
  5. Clinical findings
    - a. Recognize the clinical findings of various cardiac lesions associated with situs abnormalities and associated cardiac and extracardiac abnormalities
  6. Laboratory findings
    - a. Know what supplemental testing might be necessary in patients with situs abnormalities and how to interpret
  7. Management, including complications
    - a. Plan medical and surgical management of patients with abnormal cardiac situs
    - b. Be able to advise immunization and antibiotic prophylaxis in a patient with situs abnormalities
- F. Ectopia cordis
1. Anatomy
    - a. Recognize anatomic types and associated cardiac and noncardiac anatomic features of ectopia cordis
  2. Laboratory findings
    - a. Recognize features associated with ectopia cordis using available laboratory tests and recognize important anatomic features that could affect surgical management
  3. Management, including complications
    - a. Plan management of a patient with ectopia cordis

## 11. Disorders of the Myocardium, Pericardium, Endocardium, and Vasculature

### A. Cardiomyopathies (including systolic dysfunction, diastolic dysfunction, and hypertrophic)

1. Epidemiology and genetics of cardiomyopathy
  - a. Understand the role of protein and genetic defects in familial hypertrophic cardiomyopathy
  - b. Identify the types of mutations and inheritance pattern associated with hypertrophic cardiomyopathy
2. Anatomy
  - a. Know gross and histologic features of the specific types of cardiomyopathies (DCM, RCM, HCM, ARVC)
  - b. Know gross and histologic features and natural history of secondary cardiomyopathies
  - c. Know the morphologic changes associated with myocardial dilatation and hypertrophy
  - d. Know the cardiomyopathies specific to anthracycline +/- radiation
3. Pathophysiology
  - a. Know the effects of myocardial dilatation and hypertrophy on ventricular wall tension and stress
  - b. Know the pathophysiology of congestive heart failure, including causes and functional consequences of abnormal loading conditions and altered contractility
  - c. Know the causes of alterations in heart rate causing congestive cardiac failure
  - d. Understand the roles of calcium, cyclic AMP, catecholamine and receptor function, and other neurohumoral changes during congestive heart failure
  - e. Understand the physiology of dilated cardiomyopathy, including the effects on systemic and pulmonary vasculature and pulmonary function
  - f. Understand the effect of restrictive cardiomyopathy on systemic or pulmonary vasculature, including the effects on the liver, kidneys, and lungs
  - g. Understand the effect of hypertrophic cardiomyopathy on systemic or pulmonary vasculature, including the effects on the liver, kidneys, and lungs
  - h. Understand the role of hypertrophy in adaptation in congestive heart failure
  - i. Recognize and manage pulmonary edema
  - j. Understand the changes in adrenergic function in congestive heart failure
4. Clinical features
  - a. Know the differential diagnosis of heart failure
  - b. Know the effects of end organ congestive heart failure on systemic systems (growth, development, skin, skeletal muscle, GI, renal, hepatic, etc)
  - c. Know the principles of medical therapy for congestive heart failure, including the use of digitalis, other inotropic drugs, vasodilators, diuretics, and other therapeutic options
  - d. Understand the role and limitations of mechanical cardiovascular support
  - e. Know the pathology of cardiac viral infection as it relates to myocarditis and cardiomyopathy
  - f. Know secondary metabolic causes of cardiomyopathy
  - g. Know the major nutritional causes of cardiomyopathy, including hypocalcemia, hypercalcemia, hypocupremia, iron deficiency, and selenium deficiency

- h. Recognize the clinical features of nutritional causes of cardiomyopathy
  - i. Understand the physiologic and anatomic significance of abnormal clinical and physical findings in a patient in whom dilated, hypertrophic, or restrictive cardiomyopathy is suspected and describe the details of a general examination for this patient
  - j. Know the details of the general and cardiac examination of dilated, hypertrophic, and restrictive cardiomyopathy and understand the physiologic and anatomic significance of abnormal physical findings
  - k. Understand the natural history of the various forms of cardiomyopathy
5. Laboratory testing
- a. Know the role of cardiac MRI in the assessment of various cardiomyopathies
  - b. Know the role of noninvasive imaging in dilated, hypertrophic, and restrictive cardiomyopathy
  - c. Understand the role of invasive assessment in cardiomyopathy.
  - d. Know the cardiac catheterization features of dilated, hypertrophic, and restrictive cardiomyopathy, and relate them to physiologic and/or anatomic features of the condition
  - e. Understand the significance of ambulatory and exercise monitoring results in dilated, hypertrophic, and restrictive cardiomyopathy
  - f. Relate abnormal electrocardiographic features to physiologic and/or anatomic details of dilated, hypertrophic, or restrictive hypertrophic cardiomyopathy
  - g. Know the role of natriuretic peptide in congestive heart failure, including the utility of monitoring serum B-type natriuretic peptide
6. Management and complications
- a. Recognize risk factors for sudden cardiac death in hypertrophic cardiomyopathy
  - b. Understand the physiology of low cardiac output from primary LV systolic dysfunction
  - c. Understand the physiology of low cardiac output from primary LV diastolic dysfunction
  - d. Know the role/indications of an implantable defibrillator in a patient with dilated cardiomyopathy; know the indications for ICD in hypertrophic cardiomyopathy
  - e. Know the role of interventional and surgical management of outflow obstruction in hypertrophic cardiomyopathy
  - f. Know the role of a mechanical ventricular assist device in the management of cardiomyopathy
  - g. Know the role of pharmacologic management of cardiomyopathies
- B. Cardiomyopathy in muscular dystrophies
- 1. Understand the genetic basis, natural history, and management of the cardiovascular manifestations of the muscular dystrophies
  - 2. Understand the genetic basis, natural history, and management of the cardiovascular manifestations of Friedreich ataxia
- C. Cardiomyopathies in metabolic diseases (includes storage diseases)
- 1. Recognize and diagnose mitochondrial diseases and manage their cardiovascular manifestations
  - 2. Recognize and diagnose fatty acid oxidation disorders, and manage their cardiovascular manifestations

3. Understand the genetic basis, natural history, and management of cardiovascular manifestations of glycogen storage diseases
  4. Understand the genetic basis, natural history, and management of the cardiovascular manifestations of the mucopolysaccharidoses
  5. Understand the genetic basis, natural history, and management of the cardiovascular manifestations of the mucopolipidoses
  6. Understand the genetic basis, natural history, and management of the glycoproteinoses
  7. Understand the genetic basis, natural history, and management of the cardiovascular manifestations of the lipidoses
- D. Transplant medicine
1. Recognize indications and contraindications for cardiac transplantation in a patient with cardiomyopathy
  2. Recognize indications and contraindications for cardiac transplantation in a patient with single ventricle
  3. Understand the determinants of an acceptable donor heart
  4. Recognize and understand the mechanism of the side effects of usual immunosuppressive drugs given to cardiac transplant recipients (in particular steroids, and calcineurin inhibitors)
  5. Know special problems of infection and lymphoproliferative disease in an immunosuppressed patient who has undergone cardiac transplantation
  6. Know current 1-year and 5-year survival rates following cardiac transplantation for infants and adolescents
  7. Assess long-term outcomes and complications of cardiac transplantation
  8. Recognize the clinical and angiographic features of graft vasculopathy, including the setting in which it occurs
  9. Plan the appropriate management of transplant-related graft vasculopathy
  10. Diagnose and manage acute signs of rejection of a transplanted heart
- E. Pericardial diseases
1. Pathophysiology
    - a. Know the etiology of major types of congenital and acquired pericardial disorders
  2. Clinical and laboratory recognition
    - a. Recognize the clinical features and laboratory manifestations of postpericardiotomy syndrome
    - b. Recognize cardiac tamponade based on clinical and laboratory manifestations
    - c. Differentiate between restrictive cardiomyopathy and constrictive pericarditis
    - d. Know the role of laboratory testing in pericarditis
    - e. Know the clinical presentation of pericarditis
  3. Management, including complications
    - a. Plan appropriate medical management for a patient with pericardial disorders
    - b. Know the role of laboratory testing in pericardial disorders
    - c. Know the indications for surgical pericardial stripping procedure in a patient with constrictive pericarditis
- F. Cardiac tumors
1. Know the gross and histologic features and natural history of cardiac tumors
  2. Know the clinical presentation of cardiac tumors

3. Plan appropriate management (including genetic counseling) of a patient with cardiac tumor
  4. Know the clinical management of patients with rhabdomyoma
- G. Pulmonary hypertension
1. Formulate a differential diagnosis for pulmonary hypertension based upon history, physical examination, and testing
  2. Understand the pathophysiology of acute and chronic pulmonary hypertension
  3. Diagnose and treat primary pulmonary hypertension of the newborn
  4. Know major problems of unoperated complex cardiac disease (eg, single ventricle) in adolescents
  5. Know major problems of pulmonary vascular obstructive disease in adolescents
  6. Know how to manage unoperated complex cardiac disease (eg, single ventricle) in adolescents
  7. Know how to manage pulmonary vascular obstructive disease in adolescents
  8. Understand the pathophysiology of pulmonary hypertension secondary to congenital heart disease
  9. Understand the secondary causes of pulmonary hypertension unrelated to congenital heart disease

## 12. Heart Function and Disease in the Fetus and Newborn

- A. Fetal diagnosis and management of congenital heart disease
1. Plan the diagnosis and management of the cyanotic newborn
  2. Know the natural history of obstructive cardiac lesions, restrictive ductus arteriosus, and restrictive ASD in the fetus
  3. Interpret a fetal echocardiogram, including developing a differential diagnosis
  4. Recognize the etiology and genetic syndromes associated with congenital heart disease in the fetus
  5. Know the indications and limitations of fetal echocardiography on the diagnosis of congenital heart disease
  6. Understand the role of routine fetal ultrasonography in screening for fetal heart disease
  7. Understand the indications, limitations, and types of fetal intervention for congenital heart defects and arrhythmias
  8. Know how to counsel a family about a fetus with congenital heart disease
- B. Fetal arrhythmias
1. Recognize and manage fetal arrhythmias
  2. Know the echocardiographic / Doppler findings in a fetus that signify abnormal flow and fetal distress
  3. Recognize and manage fetal tachycardias
  4. Recognize and manage fetal bradycardia (including heart block)
  5. Recognize and manage fetal ectopy
- C. Effects of systemic disease on the fetal heart
1. Recognize chromosomal abnormalities associated with congenital heart disease in the fetus
  2. Recognize extracardiac malformations in the fetus associated with congenital heart disease
  3. Recognize and manage hydrops fetalis
  4. Recognize and manage the complications of twin-to-twin transfusion syndrome

5. Know how to recognize and manage the cardiovascular manifestations of metabolic abnormalities in a newborn infant
- D. Effects of maternal disease on the fetal heart
  1. Recognize the cardiac anomalies associated with maternal alcohol usage
  2. Recognize and manage the effects of maternal diabetes on the newborn heart
  3. Recognize the cardiac anomalies associated with maternal use of prescription and over-the-counter medications
  4. Understand the natural history of cardiac abnormalities in the infant of a diabetic mother
  5. Recognize the fetal cardiac abnormalities associated with maternal diabetes
  6. Differentiate and manage the various causes of systemic hypertension in newborn infants
  7. Recognize the fetal cardiac abnormalities in and plan follow-up for fetuses of mothers with connective tissue diseases, including systemic lupus erythematosus
- E. Transitional physiology and pulmonary hypertension of the newborn
  1. Know the effects of birth asphyxia on neonatal cardiac performance
  2. Know how to evaluate an asphyxiated newborn infant
  3. Plan the evaluation and management of a newborn infant with transient myocardial ischemia
  4. Plan the medical management of a newborn infant with persistent pulmonary hypertension, recognizing the systemic and pulmonary effects of vasoactive drugs
  5. Understand the risk factors for development of persistent pulmonary hypertension in a newborn infant
  6. Recognize the clinical features of an infant with persistent pulmonary hypertension and interpret diagnostic studies
  7. Know the cardiovascular manifestations of maternal and fetal thyroid disease in a newborn infant
- F. Initial stabilization and management of the newborn with congenital heart disease
  1. Understand and be able to apply the principles of initial stabilization and management of the newborn with congenital heart disease
- 13. Intensive Care Management of Patients with Congenital Heart Disease**
  - A. Airway support, mechanical ventilation, and effects on congenital heart disease
    1. Understand the principles of end-tidal PCO<sub>2</sub> determination in a patient with cardiovascular disease
    2. Be familiar with airway support and mechanical ventilation strategies specific to congenital heart disease physiology
    3. Recognize and manage acute respiratory distress syndrome
    4. Know how to recognize and manage hypoventilation in a child who is mechanically ventilated
    5. Be familiar with airway support and mechanical ventilation strategies specific to patients with systolic and diastolic dysfunction
  - B. Low cardiac output state
    1. Understand the principles of supply-demand balance (eg, concept of oxygen supply, oxygen reserve, extraction, oxygen consumption, perfusion requirements, relations between supply and demand)
    2. Understand the clinical and mechanical forces controlling cardiac output



3. Understand compensatory cardiovascular adaptive mechanisms (eg, autoregulation, local response, systemic responses)
4. Know the principles of monitoring and therapy for patients with low cardiac output, eg, principles and limitations of near-infrared spectrometry
- C. Cardiopulmonary resuscitation
  1. Understand the principles of CPR, including the use of automatic electrical defibrillators
  2. Understand the physiologic principles and management for the post-arrest patient
- D. Acute management of pulmonary hypertension
  1. Recognize and manage pulmonary hypertension in the postoperative period
- E. Multiorgan system management
  1. Understand the risks to the central nervous system and management of CNS dysfunction in the acutely ill CHD patient
  2. Understand the risks to the renal system and management of renal dysfunction in the acutely ill CHD patient
  3. Understand the risks to the gastrointestinal system and management of GI dysfunction in the acutely ill CHD patient
  4. Understand the pathophysiology of hematologic complications in the postoperative and acutely ill CHD patient
- F. Quality improvement
  1. Be familiar with the quality improvement process and its application in the ICU setting
  2. Be familiar with established privacy issues in an ICU setting
  3. Be familiar with reporting responsibilities for effective QI process such as reporting to supervisors
- G. Perioperative issues
  1. Know the acute postoperative management for antiplatelet and anticoagulation therapy for artificial valves, systemic to pulmonary artery shunts, conduits and pulmonary patients at risk of pulmonary or systemic thromboembolism
  2. Know how to recognize and manage postoperative pulmonary edema and its causes
  3. Recognize and manage extracardiac complications after heart surgery
  4. Recognize, and manage surgical bleeding after cardiovascular surgery
  5. Recognize and manage the physiology of postpericardiotomy syndrome
  6. Diagnose arrhythmias in the postoperative patient
  7. Recognize and manage residual cardiac lesions after surgery may adversely affect a patient's postoperative course
  8. Know the indications for and limitations of various available diagnostic tools in evaluating a postoperative patient
  9. Understand factors that influence systemic and pulmonary blood flow in a postoperative patient with an aortopulmonary shunt
  10. Understand the role and limitations of mechanical cardiovascular support in the postoperative patient

#### 14. Arrhythmias

- A. General principles
  1. Classification
    - a. Formulate a differential diagnosis of rhythm disturbances causing a narrow QRS tachycardia, a wide QRS tachycardia, and bradycardia

2. Symptoms
    - a. Identify the signs and symptoms associated with rhythm abnormalities
    - b. Evaluate a patient who has experienced a cardiac arrest
    - c. Evaluate a family with a history of sudden cardiac death
  3. Basis or cause/physiology
    - a. Understand the physiologic consequences of rhythm disturbances
    - b. Understand the mechanisms involved in the genesis of cardiac arrhythmias (eg, re-entry, automaticity, conduction block)
  4. Therapy
    - a. General principles
      1. Understand the indications for acute and chronic medical management of tachy- and bradyarrhythmias
      2. Know the mechanical methods (eg, vagal maneuvers; esophageal, external, intracardiac pacing; cardioversion) available for treatment of arrhythmias
      3. Identify the clinical features and genetic causes of diseases associated with arrhythmias (eg, LQTS, ARVD, Brugada syndrome, CPVT, cardiomyopathies, preexcitation)
    - b. Autonomic interventions
      1. Know the techniques for use of vagal maneuvers (including indications, contraindications, risks, and limitations) (eg, Valsalva, ice to face, carotid sinus massage)
      2. Know the effect of vagal maneuvers in treating cardiac arrhythmias
    - c. Temporary pacing
      1. Understand the factors associated with temporary pacing (eg, indications, contraindications, risks, and limitations)
      2. Understand the basic technical aspects of the different modalities available for temporary pacing
    - d. Cardioversion/defibrillation
      1. Understand the factors associated with cardioversion/defibrillation (eg, indications, contraindications, risks, and limitations)
      2. Understand the basic technical aspects of cardioversion/defibrillation
    - e. Implantable devices
      1. Recognize pacing mode and pacemaker malfunction by ECG
      2. Understand the factors associated with permanent pacing (eg, indications, contraindications, risks, and limitations)
      3. Understand the factors associated with an implantable cardioverter-defibrillator (eg, indications, contraindications, risks, and limitations)
      4. Understand the basic technical aspects for insertion of a permanent pacemaker or an implantable cardioverter-defibrillator
      5. Understand the factors associated with bi-ventricular resynchronization pacing (eg, indications, contraindications, risks, and limitations)
- B. Ectopy
1. Clinical recognition
    - a. Recognize clinical manifestations and significance of ectopy
  2. Basis or cause

- a. Recognize the association of ectopic atrial tachycardias with surgery for congenital heart disease
- 3. Management
  - a. Plan the evaluation and management of a patient with frequent atrial or ventricular ectopy
- C. Supraventricular arrhythmias
  - 1. Sinus tachycardia
    - a. Clinical recognition
      - 1. Recognize the clinical features of sinus tachycardia
      - 2. Differentiate sinus tachycardia by surface electrocardiographic criteria
      - 3. Recognize intracardiac electrophysiologic characteristics of sinus tachycardia
    - b. Basis or cause
      - 1. Understand the mechanisms and natural history of sinus tachycardia
      - 2. Recognize the association of sinus tachycardia with surgery for congenital heart disease (acute and long-term)
    - c. Management
      - 1. Recognize and medically manage sinus tachycardia in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
      - 2. Recognize and manage the consequences of long-term sinus tachycardia
  - 2. Ectopic atrial tachycardia
    - a. Clinical recognition
      - 1. Recognize the clinical features of ectopic atrial tachycardia
      - 2. Differentiate ectopic atrial tachycardia by surface electrocardiographic criteria
      - 3. Recognize intracardiac electrophysiologic characteristics of ectopic atrial tachycardia
    - b. Basis or cause
      - 1. Understand the mechanisms and natural history of ectopic atrial tachycardia
      - 2. Recognize the association of certain ectopic atrial tachycardias with surgery for congenital heart disease (acute and long-term)
    - c. Management
      - 1. Recognize and medically manage ectopic atrial tachycardia in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
      - 2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for ectopic atrial tachycardia
      - 3. Recognize and manage the consequences of ectopic atrial tachycardia
  - 3. Multifocal atrial tachycardia
    - a. Clinical recognition
      - 1. Recognize the clinical features of multifocal atrial tachycardia
      - 2. Differentiate multifocal atrial tachycardia by surface electrocardiographic criteria
      - 3. Recognize intracardiac electrophysiologic characteristics of multifocal atrial tachycardia
    - b. Basis or cause
      - 1. Understand the mechanisms and natural history of multifocal atrial tachycardia
    - c. Management

1. Recognize and medically manage multifocal atrial tachycardia in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for multifocal atrial tachycardia
3. Recognize and manage the consequences of multifocal atrial tachycardia
4. Atrial flutter (intra-atrial reentry)
  - a. Clinical recognition
    1. Recognize the clinical features of atrial flutter
    2. Differentiate atrial flutter by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of atrial flutter
  - b. Basis or cause
    1. Understand the mechanisms and natural history of atrial flutter
    2. Recognize the association of atrial flutter with surgery for congenital heart disease (acute and long-term)
  - c. Management
    1. Recognize and medically manage atrial flutter in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for atrial flutter
    3. Recognize and manage the consequences of atrial flutter
5. Atrial fibrillation
  - a. Clinical recognition
    1. Recognize the clinical features of atrial fibrillation
    2. Differentiate atrial fibrillation by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of atrial fibrillation
  - b. Basis or cause
    1. Understand the mechanisms and natural history of atrial fibrillation
    2. Recognize the association of atrial fibrillation with surgery for congenital heart disease (acute and long-term)
  - c. Management
    1. Recognize and medically manage atrial fibrillation in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for atrial fibrillation
    3. Recognize and manage the consequences of atrial fibrillation
6. Atrioventricular node reentry
  - a. Clinical recognition
    1. Recognize the clinical features of AV node reentry
    2. Differentiate AV node reentry by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of AV node reentry
  - b. Basis or cause
    1. Understand the mechanisms and natural history of AV node reentry
  - c. Management

1. Recognize and medically manage AV node reentry in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
  2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for AV node reentry
  3. Recognize and manage the consequences of AV node reentry
7. Junctional ectopic tachycardia
- a. Clinical recognition
    1. Recognize the clinical features of junctional ectopic tachycardia
    2. Differentiate junctional ectopic tachycardia by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of junctional ectopic tachycardia
  - b. Basis or cause
    1. Understand the mechanisms and natural history of junctional ectopic tachycardia
    2. Recognize the association of junctional ectopic tachycardia with surgery for congenital heart disease (acute and long-term)
  - c. Management
    1. Recognize and medically manage junctional ectopic tachycardia in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for the congenital type of junctional ectopic tachycardia
    3. Recognize and manage the consequences of junctional ectopic tachycardias
8. Orthodromic reentry via accessory pathway
- a. Clinical recognition
    1. Recognize the clinical features of orthodromic reentry via accessory pathway
    2. Differentiate orthodromic reentry via accessory pathway by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of orthodromic reentry via accessory pathway
  - b. Basis or cause
    1. Understand the mechanisms and natural history of orthodromic reentry via accessory pathway
  - c. Management
    1. Recognize and medically manage orthodromic reentry via accessory pathway in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for orthodromic reentry via accessory pathway
    3. Recognize and manage the consequences of orthodromic reentry via accessory pathway
9. Permanent form of junctional reciprocating tachycardia
- a. Clinical recognition

1. Recognize the clinical features of the permanent form of junctional reciprocating tachycardia
  2. Differentiate the permanent form of junctional reciprocating tachycardia by surface electrocardiographic criteria
  3. Recognize intracardiac electrophysiologic characteristics of the permanent form of junctional reciprocating tachycardia
  - b. Basis or cause
    1. Understand the mechanisms and natural history of the permanent form of junctional reciprocating tachycardia
  - c. Management
    1. Recognize and medically manage the permanent form of junctional reciprocating tachycardia in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for the permanent form of junctional reciprocating tachycardia
    3. Recognize and manage the consequences of the permanent form of junctional reciprocating tachycardia
10. Antidromic reentry via accessory pathway
- a. Clinical recognition
    1. Recognize the clinical features of antidromic reentry via accessory pathway
    2. Differentiate antidromic reentry by surface electrocardiographic criteria
    3. Recognize intracardiac electrophysiologic characteristics of antidromic reentry
  - b. Basis or cause
    1. Understand the mechanisms and natural history of antidromic reentry
  - c. Management
    1. Recognize and medically manage antidromic reentry in patients of varying ages (eg, fetus, infant, child, adolescent, young adult)
    2. Understand the factors associated with electrophysiologic study (eg, indications, contraindications, risks, and limitations) and catheter- or surgical-based ablation therapy for antidromic reentry
    3. Recognize and manage the consequences of antidromic reentry
- D. Accessory AV connection and pre-excitation syndrome
1. Clinical recognition
    - a. Recognize clinical features associated with accessory atrioventricular connection or pre-excitation syndromes
    - b. Recognize associated cardiac defects in a patient with an accessory atrioventricular connection
  2. Electrocardiographic diagnosis
    - a. Recognize the ECG features of an atrioventricular connection or pre-excitation syndrome
  3. Electrophysiologic diagnosis
    - a. Recognize characteristics of accessory atrioventricular connections or pre-excitation syndromes based on electrophysiologic studies
  4. Basis or cause

- a. Know the natural history of accessory atrioventricular connections or pre-excitation syndromes
- 5. Management
  - a. Plan the management of patients with accessory atrioventricular connections or pre-excitation syndromes
- E. Ventricular arrhythmias
  - 1. Benign ventricular ectopy
    - a. Clinical recognition
      - 1. Distinguish the clinical features of benign ventricular ectopy and distinguish from more serious ventricular arrhythmias
      - 2. Know the risk factors, clinical features, and natural history of benign ventricular ectopy associated with a structurally normal heart or congenital heart disease
      - 3. Know the differential diagnosis of benign ventricular ectopy on electrocardiogram
      - 4. Identify the specific electrocardiographic features of diseases associated with benign ventricular ectopy
    - b. Basis or cause
      - 1. Understand the mechanisms and natural history of benign ventricular ectopy
    - c. Management
      - 1. Plan the acute and chronic management of benign ventricular ectopy
  - 2. Idiopathic outflow tract ventricular ectopy
    - a. Clinical recognition
      - 1. Distinguish the clinical features of benign idiopathic outflow tract ventricular ectopy
      - 2. Know the risk factors, clinical features, and natural history of life-threatening benign idiopathic outflow tract ventricular ectopy associated with a structurally normal heart or congenital heart disease
      - 3. Know the differential diagnosis of idiopathic outflow tract ventricular ectopy on electrocardiogram
    - b. Basis or cause
      - 1. Understand the mechanisms and natural history of idiopathic outflow tract ventricular ectopy
    - c. Management
      - 1. Plan the acute and chronic management of idiopathic outflow tract ventricular ectopy and ventricular fibrillation in patients with and without surgery for congenital heart disease
  - 3. Verapamil-sensitive LV septal ventricular tachycardia
    - a. Clinical recognition
      - 1. Distinguish the clinical features of benign verapamil-sensitive LV septal ventricular tachycardia
      - 2. Know the risk factors, clinical features, and natural history of life-threatening verapamil-sensitive LV septal ventricular tachycardia associated with a structurally normal heart or congenital heart disease
      - 3. Know the differential diagnosis of verapamil-sensitive LV septal ventricular tachycardia on electrocardiogram

- b. Basis or cause
  - 1. Understand the mechanisms and natural history of verapamil-sensitive LV septal ventricular tachycardia
- c. Management
  - 1. Plan the acute and chronic management of verapamil-sensitive LV septal ventricular tachycardia in patients with and without surgery for congenital heart disease
- 4. Scar-related macroreentrant ventricular tachycardia
  - a. Clinical recognition
    - 1. Distinguish the clinical features of scar-related macroreentrant ventricular tachycardia
    - 2. Know the risk factors, clinical features, and natural history of life-threatening scar-related macroreentrant ventricular tachycardia associated with a structurally normal heart or congenital heart disease
    - 3. Know the differential diagnosis of scar-related macroreentrant ventricular tachycardia on electrocardiogram
    - 4. Identify the specific electrocardiographic features of diseases associated with life-threatening scar-related macroreentrant ventricular tachycardia
  - b. Basis or cause
    - 1. Understand the mechanisms and natural history of scar-related macroreentrant ventricular tachycardia
  - c. Management
    - 1. Plan the acute and chronic management of scar-related macroreentrant ventricular tachycardia and ventricular fibrillation in patients with and without surgery for congenital heart disease
- 5. Ventricular tachycardia in cardiomyopathy
  - a. Clinical recognition
    - 1. Know the risk factors, clinical features, and natural history of life-threatening ventricular tachycardia in cardiomyopathy associated with a structurally normal heart or congenital heart disease
    - 2. Know the differential diagnosis of ventricular tachycardia in cardiomyopathy on electrocardiogram
    - 3. Identify the specific electrocardiographic features of diseases associated with life-threatening ventricular tachycardia in cardiomyopathy
  - b. Basis or cause
    - 1. Understand the mechanisms and natural history of ventricular tachycardia in cardiomyopathy
  - c. Management
    - 1. Plan the acute and chronic management of ventricular tachycardia in cardiomyopathy in patients with and without surgery for congenital heart disease
- 6. Catecholaminergic polymorphic ventricular tachycardia
  - a. Clinical recognition
    - 1. Distinguish the clinical features of benign catecholaminergic polymorphic ventricular tachycardia



2. Know the risk factors, clinical features, and natural history of life-threatening catecholaminergic polymorphic ventricular tachycardia associated with a structurally normal heart or congenital heart disease
  3. Know the differential diagnosis of catecholaminergic polymorphic ventricular tachycardia on electrocardiogram
  4. Identify the specific electrocardiographic features of diseases associated with life-threatening catecholaminergic polymorphic ventricular tachycardia
  - b. Basis or cause
    1. Understand the mechanisms and natural history of catecholaminergic polymorphic ventricular tachycardia
  - c. Management
    1. Plan the acute and chronic management of catecholaminergic polymorphic ventricular tachycardia in patients with and without surgery for congenital heart disease
7. Arrhythmogenic RV cardiomyopathy
- a. Clinical recognition
    1. Distinguish the clinical features of arrhythmogenic RV cardiomyopathy
    2. Know the risk factors, clinical features, and natural history of life-threatening arrhythmogenic RV cardiomyopathy associated with a structurally normal heart or congenital heart disease
    3. Know the differential diagnosis of benign arrhythmogenic RV cardiomyopathy on electrocardiogram
    4. Identify the specific electrocardiographic features of diseases associated with life-threatening right ventricular cardiomyopathy
  - b. Management
    1. Plan the acute and chronic management of arrhythmogenic RV cardiomyopathy in patients with and without surgery for congenital heart disease
8. Torsade de pointe ventricular tachycardia
- a. Clinical recognition
    1. Distinguish the clinical features of torsade de pointe ventricular tachycardia
    2. Know the risk factors, clinical features, and natural history of life-threatening torsade de pointe ventricular tachycardia associated with a structurally normal heart or congenital heart disease
    3. Know the differential diagnosis of torsade de pointe ventricular tachycardia on electrocardiogram
    4. Identify the specific electrocardiographic features of diseases associated with life-threatening torsade de pointe ventricular tachycardia
  - b. Basis or cause
    1. Understand the mechanisms and natural history of torsade de pointe ventricular tachycardia
  - c. Management
    1. Plan the acute and chronic management of torsade de pointe ventricular tachycardia in patients with and without surgery for congenital heart disease
- F. Long QT syndrome and channel abnormalities (see also Section XVI)
1. Clinical recognition

- a. Recognize and plan appropriate management of long QT interval syndromes and other channel abnormalities (eg, Brugada syndrome, CPMT, ARVD)
  - b. Recognize the association between long QT interval and torsades de pointes
  - 2. Basis or cause
    - a. Recognize the genotypic-phenotypic relationships and identify the genes associated with long QT syndrome and other channel abnormalities
    - b. Recognize the therapeutic implications for the long QT syndrome genotype
    - c. Know the mode of transmission, application, and interpretation of genetic tests of inherited channelopathies
  - 3. Management
    - a. Plan the medical management of arrhythmias of inherited channelopathies
    - b. Understand the indications for implantation of an intracardiac device for inherited channelopathies
    - c. Understand the potential role of cardiac sympathectomy in management of channelopathies
  - G. Atrioventricular block
    - 1. Clinical recognition
      - a. Know causes of neonatal complete atrioventricular block (maternal SLE, complex heart disease)
      - b. Recognize clinical manifestations of atrioventricular block
    - 2. Basis or cause
      - a. Know congenital defects associated with atrioventricular block
      - b. Recognize the association of cardiac surgery with atrioventricular block
      - c. Recognize noncardiac diseases associated with atrioventricular block (eg, mitochondrial myopathy, myotonic dystrophy)
      - d. Recognize acquired cardiac diseases associated with atrioventricular block (eg, Lyme disease)
      - e. Know the natural history of atrioventricular block of various causes (eg, congenital, acquired, surgically induced)
    - 3. Management
      - a. Plan appropriate management of atrioventricular block of various causes (eg, congenital, acquired, surgically induced)
  - H. Sinus node dysfunction
    - 1. Clinical recognition
      - a. Recognize specific situations involving long QT syndrome and channel abnormalities
      - b. Recognize ECG features of sinus node dysfunction
      - c. Recognize the electrophysiologic abnormalities of sinus node dysfunction
      - d. Identify symptoms associated with sinus node dysfunction
    - 2. Basis or cause
      - a. Recognize the causes of sinus node dysfunction
    - 3. Management
      - a. Know the indication for permanent pacemaker implantation in sinus node dysfunction
- 15. Acquired Forms of Cardiac Disease**
- A. Endocarditis

1. Know the risk factors and cardiac and noncardiac lesions that have the highest risk of bacterial endocarditis
  2. Know the common microorganisms responsible for endocarditis
  3. Recognize the signs and clinical manifestations of infective endocarditis and the symptoms of bacterial endocarditis resulting in left-heart versus right-heart endocarditis
  4. Recognize the symptoms of bacterial endocarditis resulting in left-heart versus right-heart endocarditis
  5. Know the criteria for diagnosing endocarditis
  6. Identify the extracardiac manifestations and complications of endocarditis and understand their mechanism(s) of development
  7. Know the indications for surgical management in a patient with endocarditis
  8. Know the indications for and timing of prophylaxis for bacterial endocarditis
  9. Know the current status and duration of therapy of antimicrobial therapy of infective endocarditis
  10. Know the common reasons why endocarditis may yield negative results of a culture
  11. Plan the management of the complications of endocarditis
- B. Myocarditis
1. Know the infectious causes of myocarditis
  2. Know the role of cardiac catheterization and endomyocardial biopsy in diagnosis and management of myocarditis
  3. Plan appropriate treatment of myocarditis
  4. Plan appropriate treatment of the cardiac manifestations of Lyme disease
  5. Formulate the differential diagnosis of an enlarged cardiac silhouette in a febrile child
  6. Formulate the differential diagnosis of an enlarged, poorly contractile left ventricle
  7. Understand the natural history of myocarditis
  8. Know gross and histologic features of major cardiovascular inflammatory disease
  9. Recognize myocarditis cardiac manifestations of systemic cardiac disease (eg, rheumatoid arthritis, Kawasaki disease, sepsis)
  10. Recognize the clinical presentation and laboratory features of myocarditis
  11. Recognize the clinical presentation of viral myocarditis
- C. Kawasaki disease
1. Know pathologic features and clinical cardiovascular manifestations of Kawasaki disease
  2. Know the sequence and time of appearance of cardiac lesions associated with Kawasaki disease
  3. Understand the indications for and the role of diagnostic imaging in initial diagnosis and management of Kawasaki disease, including patients with atypical presentation
  4. Know the sequence and timing of noncardiac findings associated with Kawasaki disease
  5. Know current recommendations for drug treatment of acute and chronic Kawasaki disease and results of long-term sequelae
- D. Rheumatic fever and rheumatic heart disease
1. Understand the etiologic features and specific anatomic features of rheumatic fever and rheumatic heart disease
  2. Understand diagnostic testing and classification of rheumatic fever

3. Know the sequence of anatomic features and natural history of rheumatic fever
  4. Know the gross and microscopic pathology of rheumatic fever
  5. Know the effect of pathologic anatomy on physiology in a patient with rheumatic fever
  6. Recognize the major and minor manifestations of acute rheumatic fever and their significance (eg, carditis, chorea, arthritis etc)
  7. Know the natural history of valve involvement in rheumatic heart disease and the influence of prophylaxis
  8. Know how to prevent recurrence of rheumatic fever
  9. Know the currently recommended drug therapy for a patient with acute rheumatic fever with and without cardiac involvement
  10. Know the indications for intervention in a patient with rheumatic heart disease
- E. Inflammatory heart disease caused by systemic disease
1. Recognize the causes and understand treatment of inflammatory heart disease caused by systemic disease
- F. Other infectious diseases affecting the heart
1. Identify the common cardiac complications of AIDS and know the management of each
  2. Recognize the cardiac findings secondary to congenital rubella
  3. Know infectious causes of cardiovascular problems in a newborn infant
- G. Cardiac trauma
1. Recognize the significance of clinical history and physical examination in the evaluation of cardiovascular complications of cardiac trauma
  2. Recognize the causes and treatments of commotio cordis
  3. Know the role of noninvasive testing and laboratory findings in evaluation of cardiac trauma
  4. Plan appropriate management for a patient having cardiovascular trauma
- H. Coronary disease and myocardial ischemia (see also Sections X and XV.C)
1. Recognize the risk factors for and the precursors to the development of risk factors for coronary artery disease
  2. Know recommendations for prevention of coronary artery disease, including diagnostic testing and exercise
- I. Artificial valves, conduits, and hematologic issues
1. Recognize major problems associated with artificial valves and plan appropriate management
  2. Regulate anticoagulation therapy (warfarin, heparin, low molecular weight heparin) in a patient with an artificial valve or conduit, including management plan at the time of an invasive procedure
  3. Manage antiplatelet / anticoagulation for noncardiac procedures and conditions
  4. Understand the effect of damaged endothelium on thrombosis
  5. Identify clinical and laboratory manifestations of embolic clotting disorders
  6. Formulate a differential diagnosis in a patient suspected of having an embolic clotting disorder
  7. Understand the physiology of embolic clotting disorders
- 16. Genetic Disorders and Syndromes of the Cardiovascular System**
- A. Epidemiology and screening of congenital heart disease

1. Know the recurrence risk for the common congenital cardiac anomalies based upon whether the mother or father is affected (parent-of-origin effect)
  2. Know the recurrence risk for the common congenital cardiac anomalies if a sibling is affected
  3. Understand appropriate use of genetic testing in unaffected children who have a family history of cardiovascular disease if a first-degree family member is affected
  4. Understand the appropriate use of genetic testing in children with congenital heart disease and extra-cardiac abnormalities such as intellectual and developmental disability
  5. Know the major associated cardiac and noncardiac conditions of trisomy 21 and manage their cardiovascular manifestations
  6. Know the major associated cardiac and noncardiac conditions of trisomy 18 and manage their cardiovascular manifestations
  7. Know the major associated cardiac and noncardiac conditions of trisomy 13 and manage their cardiovascular manifestations
  8. Recognize the clinical signs and symptoms of the cardiovascular manifestations of monosomy X (Turner syndrome) and manage their cardiovascular manifestations
  9. Recognize and diagnose Noonan syndrome and manage its cardiac manifestations
  10. Recognize and diagnose Holt Oram syndrome and its molecular pathogenesis
  11. Recognize and diagnose LEOPARD syndrome and manage its cardiac manifestations
  12. Recognize and diagnose Kartagener (dysmotile cilia) syndrome and manage its cardiac manifestations
  13. Recognize and diagnose CHARGE association and manage its cardiovascular manifestations
  14. Recognize and diagnose Barth syndrome and manage its cardiovascular manifestations
  15. Recognize and diagnose VATER association and manage its cardiovascular manifestations
  16. Recognize and diagnose Williams syndrome and manage its cardiac manifestations
  17. Recognize and diagnose the cardiac manifestations of Rubinstein-Taybi syndrome and manage its cardiac manifestations
  18. Recognize and diagnose Alagille syndrome and manage its cardiac manifestations
  19. Recognize and diagnose syndromes with chromosome 22q11 deletion and manage their cardiovascular manifestations
  20. Recognize and diagnose Ellis-van Creveld syndrome and manage its cardiac manifestations
- B. Connective tissue diseases (including Marfan syndrome)
1. Recognize cardiovascular involvement in a patient with collagen vascular disease and plan appropriate management
  2. Recognize and diagnose Marfan and related syndromes (eg, Loeys-Dietz syndrome, congenital contractural arachnodactyly) and manage their cardiovascular manifestations
  3. Recognize and diagnose the cardiovascular manifestations of the classical and vascular forms of Ehlers-Danlos syndrome and manage their cardiovascular manifestations
  4. Recognize and diagnose hereditary hemorrhagic telangiectasia (Osler-Rendu-Weber syndrome)
- C. Hematologic diseases

1. Recognize and manage the cardiovascular manifestations of sickle cell disease
  2. Recognize and diagnose thalassemia syndromes and manage their cardiovascular manifestations
- D. Tuberous sclerosis and neurofibromatosis
1. Recognize and diagnose tuberous sclerosis and manage its cardiovascular manifestations
  2. Recognize and diagnose neurofibromatosis and manage its cardiovascular manifestations
- E. Familial atrial myxoma
1. Recognize and diagnose familial atrial myxoma and manage its cardiovascular manifestations

## 17. Congenital Heart Disease in the Adolescent and Adult

- A. Transitional care
1. Understand the importance of transitional education and timing
  2. Be able to inform patients regarding health care insurance issues related to their disease
  3. Be able to advise patients regarding access to their medical records during transition
  4. Be able to advise patients regarding family planning issues
- B. Pregnancy
1. Know how to counsel an adolescent/adult with congenital heart disease regarding contraception and pregnancy
  2. Know the cardiovascular conditions that increase risk and those that are contraindications to pregnancy
  3. Know how to manage cardiac aspects of pregnancy
  4. Manage artificial valves during pregnancy
- C. Cardiac risk of long-standing systemic disease
1. Recognize and manage chronic cyanosis in a patient with pulmonary vascular obstructive disease
  2. Recognize the changes in arrhythmia risk as patients with CHD age, and understand the basic principles of risk stratification
- D. Collateral disease associated with long-standing CHD
1. Recognize and be able to treat collateral disease associated with long-standing CHD
- E. Psychosocial issues
1. Understand appropriate employment settings for an adolescent/young adult with cardiovascular disease
  2. Identify risk-taking behaviors with magnified negative consequences in an adolescent/young adult with cardiovascular disease

## 18. Core Knowledge in Scholarly Activities

- A. Principles of Use of Biostatistics in Research
1. Types of variables
    - a. Distinguish types of variables (eg, continuous, categorical, ordinal, nominal)
    - b. Understand how the type of variable (eg, continuous, categorical, nominal) affects the choice of statistical test
  2. Distribution of data
    - a. Understand how distribution of data affects the choice of statistical test
    - b. Differentiate normal from skewed distribution of data

- c. Understand the appropriate use of the mean, median, and mode
- d. Understand the appropriate use of standard deviation
- e. Understand the appropriate use of standard error of the mean
- 3. Hypothesis testing
  - a. Distinguish the null hypothesis from an alternative hypothesis
  - b. Interpret the results of hypothesis testing
- 4. Statistical tests
  - a. Understand when to use and how to interpret the chi square test
  - b. Understand when to use and how to interpret tests comparing continuous variables between two groups (eg, t test, Mann Whitney U)
  - c. Understand when to use and how to interpret tests comparing continuous variables between three or more groups (eg, ANOVA, Kruskal-Wallis)
  - d. Understand when to use paired tests
  - e. Understand the appropriate use of parametric versus nonparametric tests
  - f. Interpret a p value
  - g. Interpret a p value when multiple comparisons have been made
  - h. Interpret a confidence interval
  - i. Identify a type I error
  - j. Identify a type II error
- 5. Measurement of association and effect
  - a. Understand how to interpret relative risk and absolute risk
  - b. Understand how to interpret odds ratio
  - c. Understand how to interpret number needed to treat or harm
  - d. Understand how to interpret hazard ratio
  - e. Understand when to use and how to interpret correlation coefficient
- 6. Regression
  - a. Understand when to use and how to interpret regression analysis (eg, linear, logistic)
  - b. Understand when to use and how to interpret survival analysis (eg, Kaplan Meier)
- 7. Diagnostic tests
  - a. Recognize the importance of an independent "gold standard" in evaluating a diagnostic test
  - b. Interpret sensitivity and specificity
  - c. Interpret positive and negative predictive values
  - d. Understand how disease prevalence affects the positive and negative predictive value of a test
  - e. Interpret a receiver operating characteristic curve
- 8. Systematic reviews and meta-analysis
  - a. Understand the purpose of a systematic review
  - b. Understand the advantages of adding a meta-analysis to a systematic review
  - c. Interpret the results of a meta-analysis
- B. Principles of Epidemiology and Clinical Research Design
  - 1. Assessment of study design, performance and analysis (internal validity)
    - a. Recognize and understand the strengths and limitations of a cohort study, case control study, and randomized controlled clinical trial
    - b. Recognize the use and limitations of surrogate endpoints

- c. Understand the use of intent-to-treat analysis
    - d. Understand how sample size affects the power of a study
  - 2. Assessment of generalizability (external validity)
    - a. Understand how nonrepresentative samples can bias results
    - b. Assess how the data source (eg, diaries, billing data, discharge diagnostic code) may affect study results
  - 3. Bias and confounding
    - a. Identify common strategies in study design to avoid or reduce bias
    - b. Identify common strategies in study design to avoid or reduce confounding
  - 4. Causation
    - a. Understand the difference between association and causation
  - 5. Incidence and prevalence
    - a. Distinguish disease incidence from disease prevalence
  - 6. Screening
    - a. Understand factors that affect the rationale for screening for a condition or disease (eg, prevalence, test accuracy, risk benefit, disease burden, presence of a presymptomatic state)
  - 7. Cost benefit, cost effectiveness, and outcomes
    - a. Interpret cost-effectiveness ratios
    - b. Distinguish costs from charges
    - c. Understand quality-adjusted life years
  - 8. Measurement
    - a. Understand the types of validity that relate to measurement (eg, face, construct, criterion, predictive, content)
    - b. Distinguish accuracy from precision
    - c. Understand when to use and how to interpret a kappa coefficient
- C. Ethics in Research
  - 1. Professionalism and misconduct in research
    - a. Identify and manage potential conflicts of interest in the funding, design, and/or execution of a research study
    - b. Identify various forms of research misconduct (eg, plagiarism, fabrication, falsification)
    - c. Know how, and to whom, to report concerns of research misconduct
  - 2. Principles of research with human subjects
    - a. Understand and contrast the functions of an Institutional Review Board and a Data Safety Monitoring Board
    - b. Recognize the types of protections in designing research that might be afforded to children and other vulnerable populations
    - c. Understand the federal regulatory definitions regarding which activities are considered research and what constitutes human subjects research
    - d. Understand the federal regulatory definition of minimal risk and apply this to research involving children
    - e. Understand the ethical considerations of study design (eg, placebo, harm of intervention, deception, flawed design)
  - 3. Principles of consent and assent
    - a. Understand what constitutes informed consent in research



- b. Distinguish between consent and assent in research involving children
- D. Quality Improvement
- 1. Design of a Project
    - a. Understand various models of quality improvement and recognize that all utilize a data-informed, iterative process using tests of change to achieve a stated aim
    - b. Understand that the aim of any quality improvement project should be specific, measurable, achievable, realistic, and time-limited
    - c. Understand strategies to optimize identification of key drivers and interventions to achieve a specific aim
    - d. Understand tools to facilitate completion of quality improvement work, including key driver diagrams and process maps
    - e. Understand each phase of a Plan-Do-Study-Act (PDSA) cycle
  - 2. Data and Measurement
    - a. Differentiate between process, outcome, and balancing measures
    - b. Interpret a run chart and identify shifts, trends, and outliers in data
    - c. Differentiate between a run chart and a control chart
    - d. Differentiate between common cause and special cause variation