THE AMERICAN BOARD OF PEDIATRICS®

# CONTENT OUTLINE

# Pediatric Pulmonology

**Subspecialty In-Training,**
**Certification, and Maintenance of**
**Certification (MOC) Examinations**

# INTRODUCTION

This document was prepared by the American Board of Pediatrics Subboard of Pediatric Pulmonology 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 Pulmonology

Each Pediatric Pulmonology 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. | Clinical Diseases | 35% | 35% |
| 2. | Evaluation/Diagnosis | 11% | 12% |
| 3. | Therapy | 8% | 9% |
| 4. | Prevention | 2% | 2% |
| 5. | Lung Growth and Development | 4% | 4% |
| 6. | Structure and Function of the Respiratory System | 5% | 5% |
| 7. | Lung Defense Mechanisms | 4.5% | 4.5% |
| 8. | Cell Biology and Biochemistry | 3.5% | 3.5% |
| 9. | Gas Exchange, Ventilation-perfusion Distribution, Acid-base | 6.5% | 6.5% |
| 10. | Respiratory Mechanics | 6% | 6% |
| 11. | Control of Breathing | 3.5% | 3.5% |
| 12. | Pulmonary Vascular Physiology | 3.5% | 3.5% |
| 13. | Mechanics of Lung Inflammation, Injury, and Repair | 2.5% | 2.5% |
| 14. | Core Knowledge in Scholarly Activities | 5% | 3% |

# Pulmonology

1. **Clinical Diseases**
   A. Disorders of the upper airways
      1. Congenital abnormalities
         a. Choanal atresia/stenosis
            1. Epidemiology
               a. Know the association between choanal atresia and other congenital defects
            2. Etiology/Genetics
            3. Pathophysiology
               a. Pathology
               b. Path mechanisms and consequences
            4. Diagnosis and clinical manifestations
               a. History
                  1. Recognize choanal atresia as a cause of cardiorespiratory failure on the first day after birth
                  2. Recognize choanal atresia as a cause of apnea, cyanosis, and respiratory distress relieved with crying in a neonate
               b. Physical examination
                  1. In assessing choanal patency, recognize the importance of assessing nasal airflow while the patient's mouth is closed
                  2. Recognize that choanal atresia or stenosis may be unilateral or bilateral
               c. Imaging
                  1. Recognize the radiographic appearance of choanal atresia on radiopaque dye studies or CT scans
               d. Other investigations
                  1. Recognize that the simplest way to establish the diagnosis of choanal atresia in infants is by attempting to pass a small catheter through each nostril
                  2. Recognize that the finding that only a catheter smaller than a #8 French can be passed through the nasal passage of an infant is consistent with the diagnosis of choanal stenosis
               e. Diagnostic criteria - NA
               f. Complications
                  1. Recognize that infants with choanal atresia are at risk for cyanosis and aspiration during feeding
            5. Therapeutic approach
               a. Recognize that endotracheal intubation is the most effective initial treatment of choanal atresia in a symptomatic infant
               b. Recognize that the definitive treatment of both membranous and bony choanal obstruction is surgical excision with prolonged placement of a Silastic tube (for weeks) to prevent recurrence
            6. Prognosis
               a. Natural history
                  1. Recognize that untreated choanal atresia may result in life- threatening apnea, cyanosis, and death

b. Prognosis with therapy

b. Craniofacial abnormalities with micrognathia
   1. Epidemiology
   2. Etiology/Genetics
   3. Pathophysiology
      a. Pathology
      b. Path mechanisms and consequences
   4. Diagnosis and clinical manifestations
      a. History
         1. Recognize the risk of obstructive sleep apnea in children with micrognathia
         2. Recognize that upper airway obstruction may occur in infants with severe micrognathia
      b. Physical examination
         1. Describe the effect of positioning on breathing in children with micrognathia
         2. Recognize the physical features of mandibular hypoplasia
      c. Other investigations
         1. Recognize the role of polysomnography in evaluating the severity of the respiratory compromise associated with micrognathia that occurs during sleep
      d. Diagnostic criteria
      e. Complications
         1. Recognize the difficulties associated with feeding in infants with severe micrognathia and cleft palate (Pierre Robin sequence)
         2. Recognize that micrognathia may cause hypoventilation and hypoxemia in the absence of apnea
   5. Prevention and therapeutic approach
      a. Prevention
      b. Therapeutic approach
         1. Recognize that management of severe micrognathia includes prone positioning, placement of a nasopharyngeal airway, and ultimately a tracheostomy for persistent, severe airway obstruction
      c. Side effects of therapy
         1. Recognize that palate repair without anterior tongue displacement is likely to cause airflow obstruction in children with micrognathia
         2. Recognize that pharyngeal flap surgery may worsen upper airway obstruction during sleep in patients with micrognathia and cleft palate
   6. Prognosis
      a. Natural history
         1. Recognize that the natural history of micrognathia involves improvement associated with mandibular growth
      b. Prognosis with therapy
         1. Recognize that resolution of airway obstruction in patients with severe micrognathia is dependent on growth

2. Recognize that assessment of readiness for decannulation of a child with severe micrognathia must include nocturnal polysomnography and assessment of the airway both awake and asleep

c. Laryngeal web
   1. Diagnosis and clinical manifestations
      a. History
      b. Physical examination
         1. Recognize the clinical manifestations of complete laryngeal web (aphonia; severe airway obstruction at birth without inspiratory airflow or stridor)
         2. Recognize the clinical presentation of partial laryngeal web (inspiratory stridor, weak/hoarse voice, respiratory distress)
      c. Imaging
      d. Pulmonary function tests
      e. Other investigations
         1. Recognize the appearance of a laryngeal web on fiberoptic laryngoscopy
         2. Know that endoscopic evaluation is the investigation of choice in patients with laryngeal web
      f. Diagnostic criteria
      g. Complications
   2. Prevention and therapeutic approach
      a. Prevention
      b. Therapeutic approach
         1. Recognize that emergency tracheostomy is required to relieve obstruction caused by a complete laryngeal web
         2. Recognize that an endotracheal tube can perforate a laryngeal web, providing transient relief of the obstruction
      c. Side effects of therapy
         1. Recognize that a laryngeal web may redevelop after perforation with an endotracheal tube once the tube is removed

d. Laryngeal cysts
   1. Epidemiology
   2. Etiology
   3. Pathophysiology
      a. Pathology
      b. Path mechanisms and consequences
   4. Diagnosis and clinical manifestations
      a. History
         1. Recognize that laryngeal cysts are included in the differential diagnosis of congenital stridor
      b. Physical examination
      c. Imaging
      d. Pulmonary function tests
      e. Other investigations
         1. Recognize the appearance of laryngeal cysts on fiberoptic laryngoscopy
      f. Diagnostic criteria

g. Complications

5. Prevention and therapeutic approach
    a. Prevention
    b. Therapeutic approach
        1. Recognize that excision is the treatment of choice for laryngeal cyst
    c. Side effects of therapy
        1. Recognize that there is a risk of recurrence of laryngeal cyst following simple needle aspiration of the contents of the cyst

e. Laryngomalacia
    1. Epidemiology
        a. Recognize that laryngomalacia is the most common cause of stridor in infants
    2. Etiology/Genetics
    3. Pathophysiology
        a. Pathology
        b. Path mechanisms and consequences
    4. Diagnosis and clinical manifestations
        a. History
            1. Recognize that the clinical presentation of laryngomalacia includes onset of stridor shortly after birth, minimal respiratory distress, positional effects, and marked reduction of noise when the infant is at rest
            2. Describe the effect of position on stridor secondary to laryngomalacia (worse in supine position)
        b. Physical examination
            1. Recognize that laryngomalacia is associated with normal voice quality and pitch
        c. Imaging
            1. Recognize that the diagnosis of laryngomalacia cannot be established on the basis of standard x-ray studies of the neck
        d. Pulmonary function tests
        e. Other investigations
            1. Recognize the role of fiberoptic laryngoscopy in establishing the diagnosis of laryngomalacia
        f. Diagnostic criteria
        g. Complications
            1. Know that symptoms of laryngomalacia can include apnea and feeding problems
    5. Prevention and therapeutic approach
        a. Prevention
        b. Therapeutic approach
            1. Recognize the indications for surgical intervention in infants with laryngomalacia
            2. Know that in most instances no therapy is required for laryngomalacia
    6. Prognosis
        a. Natural history

1. Recognize that the natural history of laryngomalacia includes resolution of symptoms without therapy in early childhood

b. Prognosis with therapy

f. Vocal cord paralysis

1. Epidemiology
2. Etiology/Genetics
   a. Recognize the association between vocal cord paralysis and Arnold-Chiari malformation
   b. Know the most common etiology of bilateral vocal cord paralysis in a neonate
3. Pathophysiology
   a. Pathology
   b. Path mechanisms and consequences
      1. Know that vocal cord paralysis can result from local trauma
      2. Know that left-sided vocal cord paresis/paralysis can result from trauma to the recurrent laryngeal nerve during birth
      3. Know that left-sided vocal cord paresis/paralysis may occur as a complication of cardiac surgery (eg, ligation of a patent ductus arteriosus)
      4. Recognize that enlargement of the pulmonary arteries can cause entrapment of the left recurrent laryngeal nerve leading to left vocal cord paralysis
4. Diagnosis and clinical manifestations
   a. History
      1. Know that hoarse voice and mild stridor are associated with unilateral vocal cord paralysis
      2. Know that bilateral vocal cord paralysis is rarely associated with abnormal vocalization
      3. Recognize that bilateral vocal cord paralysis is associated with increased risk of recurrent aspiration
      4. Recognize the clinical presentation of vocal cord paralysis
   b. Physical examination
      1. Recognize that signs of birth trauma indicate the possibility of vocal cord paralysis
      2. Recognize that vocal cord paralysis may be an early sign of brain stem or spinal cord compression
   c. Imaging
      1. Recognize the role of magnetic resonance imaging (MRI) of the upper spinal cord and brain stem in evaluating patients with unexplained bilateral vocal cord paralysis
   d. Other investigations
      1. Recognize the findings associated with unilateral or bilateral vocal cord paralysis on fiberoptic laryngoscopy
   e. Diagnostic criteria
   f. Complications
5. Prevention and therapeutic approach

a. Prevention
b. Therapeutic approach
    1. Recognize that the presence of aspiration and the degree of airway obstruction are the primary indicators of need for therapy in patients with vocal cord paralysis
    2. Recognize that temporary relief of the symptoms of vocal cord paralysis can be provided by the use of continuous positive pressure
    3. Recognize that decompression surgery is required to relieve vocal cord paralysis secondary to Arnold-Chiari malformation

6. Prognosis
    a. Know the natural history of vocal cord paralysis secondary to birth trauma and cardiac surgery

g. Subglottic stenosis
1. Epidemiology
    a. Recognize that chronic subglottic stenosis occurs in congenital and post-traumatic forms
    b. Recognize that even brief periods of intubation may result in chronic subglottic stenosis
2. Etiology/Genetics
3. Pathophysiology
    a. Pathology
    b. Path mechanisms and consequences
        1. Recognize the role of airway inflammation secondary to trauma in the pathogenesis of acquired subglottic stenosis
        2. Recognize that the cricoid cartilage, because it is a complete ring, is predisposed to traumatic injury and stenosis
4. Diagnosis and clinical manifestations
    a. History
        1. Recognize the importance of a history of recurrent croup or a protracted croup illness in identifying a population with underlying subglottic stenosis
        2. Recognize the importance of a history of previous intubation or airway instrumentation in alerting the clinician to a diagnosis of acquired chronic subglottic stenosis
    b. Physical examination
        1. Know that significant subglottic stenosis acts as a fixed upper airway obstruction and causes noisy breathing on both inspiration and expiration
        2. Recognize the relationship between the pitch of stridor and the severity of the obstruction in chronic subglottic stenosis
        3. Recognize the physical findings (retractions, flaring, high-pitched stridor, diminished air entry) associated with significant subglottic stenosis
    c. Imaging
        1. Recognize the lack of correlation between the radiographic appearance of subglottic stenosis and the actual degree of narrowing on endoscopy

2. Know the typical findings on pulmonary function studies in a patient with subglottic stenosis

d. Pulmonary function tests

e. Other investigations

1. Recognize findings typical of subglottic stenosis on endoscopy

f. Diagnostic criteria

g. Complications

1. Know that the finding of hypoxemia or carbon dioxide retention in a child with subglottic stenosis indicates a severe obstruction since these are the results of marked hypoventilation

5. Prevention and therapeutic approach

a. Prevention

1. Recognize the role of prolonged intubation, traumatic intubation, and the use of oversized endotracheal tubes in the pathogenesis of subglottic stenosis

b. Therapeutic approach

1. Recognize the indications for surgical intervention in subglottic stenosis
2. Know that a cricoid split procedure may provide an alternative to tracheostomy in infants with subglottic stenosis

c. Side effects of therapy

1. Know that tracheal stenosis may be a complication of prolonged endotracheal intubation

6. Prognosis

a. Natural history

1. Recognize that severe subglottic stenosis is unlikely to improve with age
2. Recognize that chronic subglottic stenosis is associated with increased frequency of croup-like illnesses and delayed resolution of viral croup illness

b. Prognosis with therapy

1. Recognize that children who require tracheostomy in the treatment of subglottic stenosis will most likely need reconstructive surgery prior to successful decannulation

h. Subglottic hemangioma

1. Epidemiology

a. Recognize subglottic hemangioma as a rare cause of upper airway obstruction in children

2. Etiology/Genetics

3. Diagnosis and clinical manifestations

a. History

1. Recognize that subglottic hemangiomas should be considered in the differential diagnosis of chronic upper airway obstruction

b. Physical examination

1. Recognize the association between the presence of cutaneous hemangiomas and subglottic hemangiomas in the child with stridor

c. Imaging

1. Recognize the limitations of standard radiographic techniques in identifying the presence of a subglottic hemangioma

d. Other investigations
1. Recognize the appearance of a subglottic hemangioma on fiberoptic endoscopy

e. Diagnostic criteria

f. Complications
1. Recognize that a subglottic hemangioma may increase in size, resulting in worsening of airway obstruction

4. Therapeutic approach
a. Know potential medical therapies for a clinically significant subglottic hemangioma
b. Recognize the indications for tracheostomy in a child with a subglottic hemangioma

5. Prognosis
a. Natural history
1. Recognize that an airway hemangioma is likely to shrink with age and usually does not require therapy

b. Prognosis with therapy

i. Laryngotracheoesophageal cleft
1. Epidemiology
a. Recognize that laryngotracheoesophageal cleft is a rare cause of recurrent aspiration

2. Etiology/Genetics

3. Pathophysiology
a. Pathology
b. Path mechanisms and consequences
1. Recognize that laryngotracheoesophageal cleft is a defect that involves the anterior wall of the upper esophagus and the posterior aspect of the larynx, with the defect lying in the interarytenoid space

4. Diagnosis and clinical manifestations
a. History
1. Recognize that recurrent aspiration with feeding is the most common clinical history associated with laryngotracheoesophageal cleft

b. Physical examination

c. Imaging
1. Recognize the radiographic appearance of a laryngotracheoesophageal cleft on a barium swallow

d. Other investigations
1. Know that direct laryngoscopy rather than transnasal fiberoptic bronchoscopy is the method of choice for visualizing laryngotracheoesophageal cleft

e. Complications

5. Prevention and therapeutic approach
a. Prevention
b. Therapeutic approach

1. Recognize that prevention of recurrent aspiration is essential in the management of laryngotracheoesophageal cleft

c. Side effects of therapy

6. Prognosis
   a. Natural history
   b. Prognosis with therapy
      1. Recognize that the risk of recurrent aspiration persists even after surgical closure of a laryngotracheoesophageal cleft
      2. Recognize the risks of impaired vocal cord function after surgical repair of a laryngotracheoesophageal cleft

j. Congenital abnormalities of the tongue
   1. Epidemiology
   2. Etiology/Genetics
   3. Pathophysiology
      a. Pathology
      b. Path mechanisms and consequences
   4. Diagnosis and clinical manifestations
      a. History
         1. Recognize that macroglossia predisposes patients to obstructive sleep apnea
      b. Physical examination
      c. Imaging
         1. Recognize the role of a lateral-view x-ray study of the neck in identifying the anatomic relationship between the tongue and the oropharyngeal airway
      d. Other investigations
      e. Diagnostic criteria
      f. Complications
   5. Therapeutic approach
      a. Recognize the role of prone positioning in the acute management of airway obstruction due to enlargement of the tongue
   6. Prognosis
      a. Natural history
         1. Recognize that obstruction related to macroglossia may improve with age

2. Infections
   a. Viral croup (laryngotracheobronchitis)
      1. Epidemiology
      2. Etiology/Genetics
         a. Recognize the anatomic risk factors predisposing infants to airway obstruction due to viral croup
         b. Know the common viruses that cause croup
      3. Pathophysiology
         a. Pathology
         b. Path mechanisms and consequences

1. Know that because the cricoid cartilage is a complete ring, edema of airway wall in this region causes a greater reduction in airway lumen than in areas in which cartilage rings are incomplete

4. Diagnosis and clinical manifestations
   a. History
      1. Know that a history of recurrent croup suggests an underlying anatomic airway abnormality, gastroesophageal reflux, or atopy
   b. Physical examination
      1. Recognize the effects of agitation on ventilation in infants with croup
   c. Imaging
      1. Recognize the radiographic appearance of croup
      2. Recognize the lack of correlation between radiographic findings and the severity of the obstruction in acute laryngotracheobronchitis
   d. Pulmonary function tests
   e. Other investigations
   f. Diagnostic criteria
   g. Complications
      1. Know that hypoxemia and carbon dioxide retention in a child with croup are suggestive of severe upper airway obstruction or the development of lower airway or parenchymal disease
5. Prevention and therapeutic approach
   a. Prevention
   b. Therapeutic approach
      1. Know that epinephrine by inhalation provides symptomatic relief of airway obstruction in viral croup, that its effects are transient, and that it does not affect the duration of the illness
      2. Understand the principles of helium/oxygen therapy in children with severe upper airway obstruction secondary to viral croup (ie, turbulent flow through large airway is density dependent)
      3. Know that endotracheal intubation using an endotracheal tube one size smaller than predicted tube size (based on age and weight) is the preferred method of establishing an artificial airway in patients with viral croup
      4. Recognize the indications for placing an artificial airway in a child with viral croup (eg, increased frequency of treatment with epinephrine by inhalation, hypoxemia, apparent or impending carbon dioxide retention, and fatigue)
      5. Know that in acute laryngotracheobronchitis, airway obstruction may recur within one to two hours after therapy with epinephrine by inhalation, but is unlikely to recur if the patient does well for 4 hours after racemic epinephrine
   c. Side effects of therapy
      1. Know that subglottic stenosis is a complication of intubation in patients with laryngotracheobronchitis
      2. Recognize the indications for tracheostomy in patients with viral croup
6. Prognosis

a. Natural history
    1. Know that the course of viral croup in infants younger than 1 year of age is prolonged and often improves during the day with recurrence of symptoms in the early hours of the morning
b. Prognosis with therapy

b. Epiglottitis
1. Epidemiology
    a. Know that epiglottis has become rare in children due to Haemophilus influenza vaccine
    b. Know that epiglottis is more common in the elderly and immune-compromised children than in the general population
2. Etiology
    a. Know that uncommon pathogens can cause epiglottis, such as herpes group viruses and fungal infections
3. Pathophysiology
    a. Pathology
    b. Path mechanisms and consequences
        1. Recognize that the pathology of epiglottitis involves the epiglottitis and other supraglottic structures but the subglottic space and trachea are usually spared
4. Diagnosis and clinical manifestations
    a. History
        1. Know that the onset of illness of epiglottitis can be very rapid (hours), consisting of sore throat and difficulty breathing
    b. Physical examination
        1. Know that stridor is not a prominent feature of epiglottitis
    c. Imaging
        1. Recognize the radiographic appearance of acute epiglottitis (positive thumb sign on lateral-view x-ray study of the neck)
        2. Recognize the radiographic appearance of a retro-pharyngeal abscess
    d. Pulmonary function tests
    e. Other investigations
        1. Know the association between Haemophilus influenzae and epiglottitis
    f. Diagnostic criteria
    g. Complications
5. Prevention and therapeutic approach
    a. Prevention
        1. Know that haemophilus influenzae vaccination has reduced the prevalence of epiglottitis through both individual and herd immunity
    b. Therapeutic approach
        1. Know that patients with acute epiglottitis should undergo endotracheal intubation to ensure an adequate airway until inflammation subsides
        2. Know that a skilled provider needs to remain with a patient with epiglottitis until the airway is visualized and secured
    c. Side effects of therapy
6. Prognosis

a. Natural history
b. Prognosis with therapy

c. Bacterial tracheitis
1. Epidemiology
a. Know that bacterial tracheitis can be sporadic or epidemic
2. Etiology/Genetics
a. Know common causes of bacterial tracheitis
3. Pathophysiology
a. Pathology
b. Path mechanisms and consequences
4. Diagnosis and clinical manifestations
a. History
1. Know that children with bacterial tracheitis usually have a prodrome similar to that of viral croup and that high fever and severe airway obstruction subsequently develop
2. Recognize the clinical picture of bacterial tracheitis with moderate to severe respiratory distress punctuated by episodes of complete or near complete airway obstruction
b. Physical examination
1. Recognize the physical findings associated with tracheal obstruction
2. Recognize the diagnostic significance of purulent materials in the airway visible upon initial intubation
c. Imaging
1. Recognize the radiographic appearance of bacterial tracheitis
d. Pulmonary function tests
e. Other investigations
1. Know the role of bronchoscopy in establishing the diagnosis of bacterial tracheitis
f. Diagnostic criteria
g. Complications
5. Prevention and therapeutic approach
a. Prevention
b. Therapeutic approach
c. Side effects of therapy
1. Know that pulmonary edema is a complication of relief of airway obstruction
6. Prognosis
a. Natural history
1. Recognize that intubation may be required for a prolonged period and that even with intubation, sudden and severe airway obstruction can develop, necessitating suctioning and reintubation
b. Prognosis with therapy
1. Know that delay in treatment of bacterial tracheitis can result in death

B. Congenital disorders of the lower airway
1. Epidemiology

a. Know that congenital pulmonary lymphangiectasia may be primary and associated with generalized lymphangiectasia or syndromes such as Noonan syndrome and anatomic abnormalities such as pulmonary vein stenosis or lymphatic obstruction

2. Etiology/genetics
   a. Know that unilateral lung agenesis most likely represents defective development during the embryonic stage
   b. Know that in most cases of congenital lobar emphysema a cause cannot be found, but identified etiologies may include central and lung hyperplasia
   c. Know that congenital diaphragmatic hernia involves an abnormality of early embryologic development
   d. Know that sequestration involves an interruption in orderly early lung development and a persistence of the primitive perfusion of the sequestered lung tissue from the systemic circulation
   e. Know that absent fetal breathing, oligohydramnios, or diaphragmatic hernia may be associated with lung hypoplasia
   f. Know that tracheomalacia may be idiopathic or associated with bronchopulmonary dysplasia, tracheoesophageal fistula, vascular ring, or other lesions
   g. Know that a bronchogenic cyst represents an abnormality of early airway development
3. Pathophysiology
   a. Pathology
      1. Know that characteristic histologic findings in congenital lobar emphysema vary but may include decreased bronchial wall cartilage
      2. Know that congenital lobar emphysema rarely involves the lower lobes
      3. Know the usual sites of diaphragmatic hernias
      4. Know the anatomic characteristics of intralobar and extralobar sequestrations
      5. Know the common location of pulmonary sequestration (ie, left-sided, lower lobe, posterior, and medial)
      6. Know that sequestered lobes are usually perfused by an abnormal systemic artery that may arise from below the diaphragm
      7. Know which congenital malformations are supplied by a systemic artery
      8. Know that hypoplastic lungs are initially small, with decreased numbers of alveoli and decreased caliber of airways and vessels
      9. Recognize the association between lung hypoplasia and the other manifestations of Potter syndrome
      10. Know that a tracheal bronchus most commonly arises on the right from the midintrathoracic region of the trachea
      11. Recognize the patterns of tracheoesophageal fistula
      12. Recognize the association between right or double aortic arch and compression of the trachea due to the vascular ring that is made up in part by these vessels
      13. Know that most closed vascular rings include the esophagus and trachea, while pulmonary sling is posterior to the trachea but anterior to the esophagus
      14. Know that the scimitar syndrome includes abnormal pulmonary venous drainage and hypoplastic lung
      15. Know the usual anatomic location of bronchogenic cysts in the middle or posterior mediastinum

16. Know that neurenteric cysts are associated with vertebral abnormalities
17. Know that a negative neutrophil oxidative burst test is associated with chronic granulomatous disease

b. Path mechanisms and consequences
   1. Know that lobes involved in congenital lobar emphysema and cystic adenomatoid malformations may compromise respiratory reserve by compressing uninvolved lobes
   2. Know that congenital diaphragmatic hernia is usually associated with hypoplasia of the ipsilateral and, to a lesser extent, of the contralateral lung
   3. Know that in individuals with congenital diaphragmatic hernia, respiratory compromise at birth is exacerbated by bowel distention
   4. Know that limited gas exchange capability and increased pulmonary vascular resistance are primary physiologic abnormalities in pulmonary hypoplasia
   5. Know that aspiration is the primary cause of lung injury in patients with tracheoesophageal fistula
   6. Know that patients with extrinsic compression of the trachea (as in vascular ring) characteristically have relatively normal oxygenation despite significant respiratory distress
   7. Know that central airway obstruction can be caused by a complete tracheal rings
   8. Know that tracheomalacia is associated with dynamic collapse of the trachea
   9. Know why airway obstruction due to tracheomalacia commonly worsens with disease causing peripheral airway obstruction such as bronchiolitis

4. Diagnosis and clinical manifestations
   a. History
      1. Know that congenital lesions that occupy intrathoracic space may manifest as respiratory insufficiency (newborn); as respiratory distress w/minor respiratory infections (1st yr); or as asymptomatic findings on x-ray study of the chest
      2. Know that congenital diaphragmatic hernia is usually symptomatic at birth but can be an unexpected finding on x-ray study of the chest later in life
      3. Know that sequestered lobes are usually asymptomatic until infected
      4. Know that recurrent "pneumonia" in the same lobe with persistently abnormal findings on x-ray study of the chest are suggestive of a pulmonary sequestration
      5. Know that cystic adenomatoid malformation commonly becomes symptomatic in the newborn period or early infancy
      6. Know that an individual with a vascular ring may have dysphagia
   b. Physical examination
      1. Know that the chest may be small in individuals with lung hypoplasia
   c. Imaging
      1. Recognize findings typical of pulmonary sequestration on chest radiograph and computed tomography of the chest
      2. Recognize findings typical of congenital lobar emphysema on chest radiograph and computed tomography of the chest
      3. Recognize findings typical of cystic adenomatoid malformation on chest radiograph and computed tomography of the chest

4. Recognize findings typical of congenital diaphragmatic hernia on chest radiograph
5. Recognize findings typical of bronchogenic cyst on chest radiograph and computed tomography of the chest
6. Recognize findings typical of hypoplastic lung on chest radiograph and computed tomography of the chest
7. Know that in congenital lobar emphysema and cystic adenomatoid malformation, affected lobes may remain filled with fluid and radiodense longer than unaffected lobes after birth
8. Know that the appearance of congenital diaphragmatic hernia may be difficult to differentiate from that of cystic adenomatoid malformation on a plain chest radiograph
9. Know the imaging techniques that should be used to diagnose vascular ring and their interpretation
10. Recognize the usual radiographic findings in the scimitar syndrome
11. Know that tracheomalacia is best diagnosed by studies that show dynamic collapse of the trachea
12. Know the danger of positive pressure ventilation in an infant with congenital lobar emphysema
13. Recognize the findings of H-type tracheoesophageal fistula on imaging studies

d. Pulmonary function tests
e. Other investigations
    1. Recognize the bronchoscopic findings typical of vascular ring
    2. Recognize findings typical of bronchomalacia on bronchoscopy
    3. Recognize the bronchoscopic findings typical of H-type tracheoesophageal fistula
    4. Recognize the difficulty of diagnosing tracheomalacia or bronchomalacia by means of bronchoscopy in the absence of spontaneous ventilation
    5. Recognize findings typical of tracheomalacia on bronchoscopy
    6. Recognize the difficulty of diagnosing tracheomalacia by means of rigid bronchoscopy with positive pressure ventilation
f. Diagnostic criteria
g. Complications
    1. Know that in congenital lobar emphysema, affected lobes may increase in volume acutely with positive pressure ventilation
    2. Know that recurrent infection is a common complication of cystic adenomatoid malformation, bronchogenic cysts, and sequestered lobes
    3. Know that there is a small but finite incidence of malignancies arising in congenital cystic adenomatoid malformations

5. Prevention and therapeutic approach
    a. Prevention
    b. Therapeutic approach
        1. Know that surgical excision is indicated for congenital lobar emphysema when respiratory reserve is compromised
        2. Know that both continuous positive airway pressure and sedation may improve airflow in patients with tracheomalacia

3. Know that both continuous positive airway pressure and sedation may improve airflow in patients with bronchomalacia

c. Side effects of therapy

6. Prognosis

a. Natural history

1. Know that lobes affected by congenital lobar emphysema usually remain approximately the same size after the first year of age, and therefore become smaller in relation to the remainder of the normal lung
2. Know that the natural histories of sequestration, cystic adenomatoid malformation, and bronchogenic cyst include recurrent infection and abscess formation once initial infection has occurred

b. Prognosis with therapy

1. Know that after surgery for congenital diaphragmatic hernia, lung volume may return to normal, but pulmonary perfusion and respiratory reserve are usually permanently abnormal
2. Know that the unaffected lobes are usually normal in patients with cystic adenomatoid malformation, bronchogenic cyst, and sequestration
3. Know that local tracheomalacia and a brassy cough are common after correction of tracheoesophageal fistula
4. Know that esophageal dysmotility and recurrent aspiration remain after repair of tracheoesophageal fistula
5. Know that individuals with vascular ring may have localized tracheal narrowing before and after corrective surgery
6. Know that tracheoesophageal fistula may recur after surgical correction
7. Know that individuals with vascular ring may have tracheomalacia before and after corrective surgery

C. Asthma

1. Epidemiology

a. Definition

1. Know that asthma is defined as a complex disorder characterized by variable and recurring symptoms, airflow obstruction with bronchial hyperresponsiveness, and underlying airway inflammation
2. Know that asthma is the most common chronic lung disease in children, affecting 5% to 15% of children
3. Understand the influence of gender on the prevalence of asthma (i.e., that in prepubertal children asthma is more common in males while in postpubertal children it is more common in females)
4. Know that most infants who wheeze in early infancy are asymptomatic by 6 years of age
5. Know that Puerto Rican and non-Hispanic black children have higher asthma prevalence rates compared to non-Hispanic white children
6. Know that Puerto Rican and non-Hispanic black children have higher asthma morbidity and mortality compared to non-Hispanic white children

b. Risk factors

1. Understand the relationship between viral infection and asthma

2. Understand that allergic factors predispose the majority of patients to asthma, but also not all asthmatic patients have allergy
3. Know that maternal smoking is associated with increased wheezing in infants
4. Recognize the socioeconomic and psychosocial factors that are associated with increased morbidity and mortality in patients with asthma
5. Recognize the factors in the medical history of a child with asthma that are associated with increased morbidity and mortality, (e.g., previous severe exacerbation with intubation or ICU admission; frequent hospitalizations or ED visits)
6. Know that environmental tobacco smoke exposure is an important contributing factor for triggering asthma symptoms

2. Etiology/genetics
   a. Know that asthma, allergic rhinitis, and eczema are embraced under the general term atopic hypersensitivity and that the genetics of atopic hypersensitivity is complex
   b. Know that asthma, occurring within or outside the context of atopy, is a complex genetic disorder with a heterogeneous phenotype, largely attributed to the interactions among many genes and between these genes and the environment
3. Pathophysiology
   a. Pathology
      1. Recognize the histopathologic features of chronic asthma
      2. Recognize the histopathologic features of status asthmaticus
   b. Pathogenic mechanisms and consequences
      1. Know the noninfectious agents that exacerbate bronchial hyperresponsiveness and asthma symptoms
      2. Know the infectious agents that exacerbate bronchial hyper- responsiveness and asthma symptoms
      3. Understand that the degree of airway hyperreactivity to histamine or methacholine correlates with the number of mast cells, eosinophils, and desquamated epithelial cells detected on bronchoalveolar lavage
      4. Understand the role of the parasympathetic nervous system in asthma
      5. Understand the role of the sympathetic nervous system in asthma
      6. Know that stimulation of the nonadrenergic, noncholinergic nervous system can lead to neurogenic inflammation in patients with asthma
      7. Know the agents that are released from lung mast cells that may play a role in asthma
      8. Understand the mechanism involved in the early phase of airflow reduction during an asthmatic response to an inhaled-allergen challenge
      9. Understand the mechanism involved in the late phase of airflow reduction during an asthmatic response to an inhaled-allergen challenge
      10. Know the role of leukotrienes in asthma
      11. Know the role of cyclooxygenase products in asthma
      12. Understand the role of bronchial epithelium in the pathogenesis of asthma
      13. Understand the mechanisms that promote increased lung water and solute movement during status asthmaticus

14. Know that increased biomarkers of eosinophilic airway inflammation, such as eosinophil major basic protein and exhaled nitric oxide correlate with the degree of bronchial hyperresponsiveness
15. Understand the role of lymphocytes and their products in the pathogenesis of asthma

4. Diagnosis and clinical manifestations
   a. History
      1. Know the conditions that cause symptoms and findings that may be confused with asthma (eg, laryngomalacia, tracheomalacia, laryngeal web, tracheostenosis, and vascular rings)
      2. Understand that the possibility of foreign body aspiration must be considered in a young patient who has an abrupt onset of wheezing
      3. Know that cough may be the only symptom of asthma in childhood
      4. Know that the use of aspirin may induce asthma in the context of the aspirin triad (nasal polyposis, asthma, and aspirin hypersensitivity)
      5. Know the history that suggests that allergy is contributing to asthma
      6. Know the history and findings that suggest that gastroesophageal reflux is contributing to asthma
      7. Know that chronic sinus disease is associated with nocturnal cough and worsening of asthma symptoms
      8. Know that asthma can accompany other forms of obstructive lung disease
      9. Recognize the clinical presentation of exercise-induced asthma
      10. Recognize the clinical presentation of vocal cord dysfunction mimicking and/or complicating asthma
   b. Physical examination
      1. Know that the use of accessory muscles of respiration indicates a moderate to severe acute asthma attack
      2. Know that clubbing is rarely seen in patients with asthma
      3. Know the definition and significance of pulsus paradoxus in patients with asthma
      4. Know that the absence of wheezing in an acute asthma exacerbation with increased work of breathing indicates moderate to severe airway obstruction
      5. Describe the physical findings of allergy in an asthmatic patient
   c. Imaging
      1. Recognize the indications for x-ray study of the chest in a patient with acute asthma
      2. Recognize typical findings on an x-ray study of the chest in a patient with asthma
   d. Pulmonary function tests
      1. Know the changes in pulmonary function during recovery from status asthmaticus
      2. Recognize the changes in pulmonary function associated with acute asthma
      3. Understand that lung function in a child with mild asthma is usually normal between attacks
      4. Know that mean pleural pressure is markedly subatmospheric during inspiration in patients with status asthmaticus

5. Understand the relation between (PaO2 and PaCO2) and changes in spirometry in asthma
6. Recognize the patterns of PaCO2 and PaO2 values in an acute asthma attack that progresses to respiratory failure
7. Understand the mechanisms that lead to hypoxemia in asthma
8. Understand the mechanisms that lead to alteration of PaCO2 in acute asthma
9. Understand the utility and limitations of a peak flow meter in the management of asthma
10. Understand that significant variation between morning and evening peak flow rates indicates worsening asthma
11. Know that an adequate exercise challenge test requires near maximal exercise for at least 6 minutes
12. Understand the time sequence of bronchodilation and bronchoconstriction during exercise in asthmatic children
13. Know the indications and specificity of the methacholine challenge in the diagnosis of asthma
14. Know that bronchial provocation tests can normalize with treatment in children with asthma
15. Understand that lung function in a child with moderate asthma commonly demonstrates normal FVC and FEV1 values, but a diminished FEV1/FVC ratio

e. Other investigations
1. Recognize the general laboratory findings characteristic of an atopic child
2. Understand the usefulness and limitations of radioallergosorbent tests and IgE measurements in the assessment of asthmatic patients
3. Understand that allergic bronchopulmonary aspergillosis is associated with a markedly increased serum IgE concentration

f. Diagnostic criteria
1. Know the criteria for a positive exercise test for exercise-induced asthma; i.e. a 10% decline in FEV1
2. Know how to interpret PC20 data from a methacholine challenge

g. Complications
1. Know that lobar atelectasis is a complication of asthma
2. Know that pneumothorax is a life-threatening complication of acute asthma
3. Know the contraindications for bronchial provocation tests
4. Understand how allergic bronchopulmonary aspergillosis can complicate asthma
5. Know the diagnostic criteria for allergic bronchopulmonary aspergillosis
6. Know the therapy for allergic bronchopulmonary aspergillosis
7. Know the clinical and laboratory manifestations of allergic bronchopulmonary aspergillosis

5. Prevention and therapeutic approach

a. Prevention
1. Understand the importance of allergen identification and avoidance in patients with asthma
2. Understand the importance of eliminating smoke from the environment of a patient with asthma

3. Know the role of asthma education programs in the management of asthma

b. Therapeutic approach
   1. Know that theophylline is an alternative but not preferred controller medication in persistent asthma
   2. Know the indications and therapeutic guidelines for using short-acting beta-agonists in asthma
   3. Know the indications and therapeutic guidelines for using anticholinergic drugs in the treatment of acute asthma
   4. Understand the management of lobar collapse in asthma
   5. Know the benefits of systemic administration of corticosteroids to patients with asthma
   6. Know the time required for the benefits of systemic administration of corticosteroids to appear
   7. Know what conditions requiring antibiotic therapy can complicate acute asthma
   8. Know the management of pneumothorax in acute asthma
   9. Recognize the indications for oxygen therapy in a patient with acute asthma
   10. Understand the indications for and role of mechanical ventilation in the treatment of acute asthma
   11. Know the therapeutic guidelines for fluid management in acute asthma
   12. Recognize the clinical effects of cromolyn sodium on the dual asthmatic response
   13. Know that short-acting beta-2 agonists do not block the late asthmatic response
   14. Know proposed mechanisms of action of corticosteroids in patients with asthma
   15. Recognize the effect of corticosteroids on the immediate and late asthmatic response
   16. Know the therapy for exercise-induced asthma
   17. Know the relative glucocorticoid and mineralocorticoid potencies of the steroid preparations available for the treatment of asthma
   18. Recognize that metered-dose inhalers and wet nebulization are equally effective techniques in the delivery of drugs to patients with mild to moderate asthma
   19. Know the efficacy of different techniques of administering inhaled asthma medications
   20. Know that aspirin, beta-blocker, and cholinergic agonists should be used with caution in children with asthma
   21. Know that intravenous beta-agonists have not been consistently shown to be more effective than inhaled beta agonists in acute asthma
   22. Know that corticosteroids by inhalation can reduce dependence on systemic corticosteroids in patients with chronic asthma
   23. Know the types of medications that may decrease airway responsiveness in patients with asthma
   24. Know that cromolyn sodium and nedocromil are alternative, not preferred, medications for treatment of mild persistent asthma and that they can be used as preventative treatment before exercise or unavoidable exposure to known allergens
   25. Know that the use of a home peak-flow meter can be an effective adjunct to a total therapy program for asthma

26. Know the appropriate management of fluids in children with asthma
27. Recognize how to assess adherence with metered-dose inhaler therapy
28. Know the proposed mechanisms of actions for leukotriene modifiers in patients with asthma
29. Recognize that the treatment for uncomplicated pneumomediastinum consists of treating the underlying cause, rest, analgesics, and simple clinical monitoring
30. Know the indications and therapeutic guidelines for using combination drugs containing beta-agonists and long-acting corticosteroids in acute asthma
31. Know that long-acting beta-agonist monotherapy is contraindicated in children with asthma
32. Know that inhaled corticosteroids are the preferred controller medications used in persistent asthma
33. Know the relative potency of common inhaled corticosteroids
34. Know that inhaled corticosteroid dose should be increased in a stepwise manner with increased impairment (symptoms/lung function) and/or risk
35. Know that the dose response curve for inhaled corticosteroids reaches a plateau at moderate doses in the majority of patients

c. Side effects of therapy
   1. Know that administration of systemic beta-agonists is associated with more side effects than inhaled beta-agonists
   2. Understand the risks of mechanical ventilation associated with the treatment of severe acute asthma
   3. Know the manifestations of adrenal suppression in asthmatic patients being treated with inhaled and systemic corticosteroids
   4. Recognize the side effects of long-term treatment of asthma with oral corticosteroids
   5. Recognize that asthmatic children given high-dose inhaled corticosteroids may require systemic corticosteroids during stress
   6. Recognize the side effects of long-term treatment of asthma with corticosteroids by inhalation
   7. Know the side effects of anticholinergic drugs in the management of acute asthma
   8. Know the side effects of theophylline in the management of acute asthma
   9. Know the side effects of intravenous beta-agonists in the management of acute asthma
   10. Know the side effects of inhaled beta-agonists in the management of acute asthma
   11. Know the potential drug interactions in the treatment of acute asthma
   12. Understand the factors that contribute to atelectasis in patients with asthma (eg, airway plugging, oxygen therapy, muscle paralysis, endotracheal intubation)

6. Prognosis
   a. Natural history
      1. Recognize that in patients with asthma, symptoms of asthma may decrease while airway reactivity remains increased
      2. Know the relationship between age of onset and severity of childhood asthma
      3. Know the mortality rate and predictors of mortality in children with asthma

4. Know the changes in blood gases and pulmonary function measurements during the resolution of status asthmaticus
5. Know the factors that impact on the prognosis for wheezing in early infancy (i.e., risk factors for developing persistent asthma)

D. Bronchiolitis
1. Epidemiology
   a. Know that the majority of infants are infected with respiratory syncytial virus in the first year after birth
   b. Know that young age, premature birth, and a history of apnea of prematurity are risk factors for the development of apnea with viral bronchiolitis
   c. Know that certain ethnic groups are at increased risk for the development of permanent sequelae of adenoviral bronchiolitis
   d. Know that natural immunity to respiratory syncytial virus is imperfect and that reinfection is common
   e. Know that respiratory syncytial virus is transmitted by direct inoculation of virus onto the nasal or ocular mucous membrane rather than by exposure to airborne virus
   f. Know the factors associated with an increased risk of bronchiolitis
   g. Know the indications for respiratory syncytial virus immunoprophylaxis
2. Etiology/genetics
   a. Know that respiratory syncytial virus is the most common cause of bronchiolitis in infancy
   b. Know that human metapneumovirus is a pathogen that causes bronchiolitis
3. Pathophysiology
   a. Pathology
      1. Recognize the pathologic findings associated with infectious bronchiolitis
      2. Know the functional characteristics of the lung and chest wall that predispose the infant with bronchiolitis to increased morbidity
   b. Physical mechanisms and consequences
      1. Know that hypoxemia associated with bronchiolitis usually represents disruption of the normal matching of ventilation and perfusion
4. Diagnosis and clinical manifestations
   a. History
   b. Physical examination
      1. Recognize the physical findings typical of bronchiolitis
   c. Imaging
      1. Recognize radiologic findings typical of acute bronchiolitis
   d. Pulmonary function tests
   e. Other investigations
      1. Know that carbon dioxide retention is usually a late finding in patients with viral bronchiolitis
      2. Know that bacterial superinfection in the first three or four days of infection is uncommon in infants with respiratory syncytial virus bronchiolitis
      3. Know the diagnostic tests for the identification of the organism causing viral bronchiolitis
   f. Diagnostic criteria

g. Complications
    1. Know the association between bronchiolitis in infancy and subsequent lower respiratory tract symptoms
    2. Know there is increased risk of aspiration in otherwise healthy infants with respiratory syncytial virus bronchiolitis
5. Prevention and therapeutic approach
    a. Prevention
        1. Know how to reduce nosocomial transmission of respiratory syncytial virus
    b. Therapeutic approach
        1. Know the indications for oxygen therapy in infants with bronchiolitis
        2. Know the effects of adrenergic agonists on patients with viral bronchiolitis
        3. Know that antibiotic and corticosteroid therapies are rarely indicated for infants with uncomplicated viral bronchiolitis
        4. Know the treatment of pulmonary graft-versus-host disease in a patient who has received a bone marrow transplant
    c. Side effects of therapy
6. Prognosis
    a. Natural history
    b. Prognosis with therapy

E. Cystic fibrosis
1. Epidemiology
2. Etiology/genetics
    a. Given an incidence figure for cystic fibrosis in the population, be able to calculate the probable carrier rate
    b. Given the scenario of two parents who have a child with cystic fibrosis, be able to calculate the odds that future children will have cystic fibrosis, be heterozygote carriers, or be homozygous normal
    c. Be able to calculate the odds that siblings, aunts, and uncles of patients with cystic fibrosis will have a child with cystic fibrosis
    d. Know the most common gene mutation that causes cystic fibrosis in North America
    e. Recognize the benefits and limitations of newborn screening for cystic fibrosis
    f. Know the commonly used methods for cystic fibrosis newborn screening (IRT and DNA testing)
    g. Be able to interpret the results of newborn screening for cystic fibrosis and understand the need for appropriate confirmatory testing
3. Pathophysiology
    a. Pathology
        1. Know the major pathologic features of the lungs and pancreas in patients with cystic fibrosis
        2. Know the evolution of airway damage in the lungs of patients with cystic fibrosis
    b. Path mechanisms and consequences
        1. Know that chloride permeability of epithelial cells is abnormal in cystic fibrosis
        2. Know the putative relationship between the chloride defect in cystic fibrosis and abnormalities in the respiratory tract, pancreatic ducts, and sweat glands

3. Know that ion transport defect does not recur in lungs transplanted into patients with cystic fibrosis
4. Know the relationship between ion transport defects in cystic fibrosis and abnormalities in transepithelial voltage
5. Know the sequence of the functional changes in the lungs of patients with cystic fibrosis
6. Know the mechanism of hepatobiliary disease in patients with cystic fibrosis
7. Know the pathogenic features of cystic fibrosis-related diabetes mellitus and that it is distinct from either type 1 or type 2 diabetes
8. Know the mechanism of steatorrhea in patients with cystic fibrosis
9. Know the mechanism of fat-soluble vitamin deficiencies in patients with cystic fibrosis
10. Know the mechanism of hypochloremic alkalosis in infants with cystic fibrosis
11. Know the association between pancreatitis and cystic fibrosis
12. Know the signs and symptoms associated with distal intestinal obstruction syndrome in patients with cystic fibrosis
13. Know the primary bacterial pathogens associated with endobronchial infection in cystic fibrosis and understand the significance of each organism's presence in respiratory tract cultures
14. Understand the potential sources of acquisition of pathogenic bacteria in cystic fibrosis patients, including contact with other cystic fibrosis patients
15. Recognize the role of intercurrent viral respiratory tract infections in the clinical course of cystic fibrosis patients
16. Identify the primary features of inflammation in cystic fibrosis lung disease and the role of infectious pathogens in exacerbating airway inflammation in cystic fibrosis

4. Diagnosis and clinical manifestations
   a. History
      1. Recognize the clinical manifestations of meconium ileus in a newborn infant
      2. Recognize the clinical manifestations of distal intestinal obstruction syndrome
      3. Recognize the clinical manifestations of cystic fibrosis in a patient with untreated steatorrhea
      4. Recognize the clinical manifestations of a typical exacerbation of cystic fibrosis lung disease
      5. Recognize the clinical manifestations of deficiencies of the following vitamins: A, D, E, and K
      6. Recognize the clinical manifestations of cystic fibrosis-related diabetes
      7. Recognize the clinical manifestations of hepatobiliary disease in a patient with cystic fibrosis
      8. Recognize the clinical manifestation of pancreatitis in a patient with cystic fibrosis
      9. Know that extracellular DNA and F-actin contribute to the viscoelastic properties of airway secretions in cystic fibrosis
   b. Physical examination
      1. Recognize the physical findings typically associated with diffuse bronchiectasis
      2. Be able to recognize nasal polyps

3. Be able to recognize digital clubbing

c. Imaging
   1. Recognize findings of cystic fibrosis on x-ray study of the chest
   2. Recognize typical manifestations of cystic fibrosis on computed tomography (CT scan) of the chest
   3. Know that findings on x-ray studies of the sinuses are not usually helpful in the management of sinus disease in patients with cystic fibrosis
   4. Recognize the indications for bronchial arteriography in a patient with cystic fibrosis
   5. Recognize radiographic findings typical of intestinal obstruction caused by distal intestinal obstruction syndrome in a patient with cystic fibrosis

d. Pulmonary function tests
   1. Recognize pulmonary function findings typical of cystic fibrosis and its progression

e. Other investigations
   1. Know how to assess pancreatic exocrine deficiency
   2. Know the major laboratory criteria for allergic bronchopulmonary aspergillosis in patients with cystic fibrosis
   3. Recognize the typical blood gas findings associated with various stages of cystic fibrosis lung disease
   4. Recognize that both mucoid and non-mucoid strains of Pseudomonas aeruginosa may be cultured from sputum in patients with cystic fibrosis and that the mucoid strain is secondary to release of exopolysaccharide
   5. Appreciate the investigations required prior to liver transplant in cystic fibrosis
   6. Know the appropriate approach to screening for and confirmation of cystic fibrosis-related diabetes
   7. Understand appropriate physical and laboratory methods for detection of malnutrition in cystic fibrosis patients

f. Diagnostic criteria
   1. Understand the rationale for a minimum volume or weight of sweat required for an acceptable sweat test
   2. Know that a single sweat test result by itself does not confirm or negate the diagnosis of cystic fibrosis
   3. Recognize the indications for repeating a sweat chloride measurement
   4. Recognize the value and limitations of genetic testing in the diagnosis of cystic fibrosis
   5. Know the diagnostic criteria for cystic fibrosis

g. Complications
   1. Recognize the association between pulmonary hemorrhage and cystic fibrosis
   2. Know that pneumothorax is a complication of cystic fibrosis lung disease
   3. Know that nasal polyposis is a frequent complication of cystic fibrosis
   4. Know that sterility is very common in males with cystic fibrosis
   5. Know that portal hypertension may result from hepatobiliary disease in patients with cystic fibrosis
   6. Know that dehydration and hypotension may be the initial symptoms in a patient with cystic fibrosis

7. Recognize that the incidence of cystic fibrosis-related diabetes increases with increasing age in patients with cystic fibrosis
8. Recognize that cor pulmonale is a complication of advanced cystic fibrosis lung disease
9. Recognize the complications associated with advanced pulmonary disease in patients with cystic fibrosis
10. Know that massive hemoptysis in a patient with cystic fibrosis usually originates from the bronchial circulation

5. Prevention and therapeutic approach
   a. Prevention
      1. Know that prenatal diagnosis of cystic fibrosis is possible
      2. Be able to interpret CFTR genetic testing results from prospective parents who are screened prior to conception
   b. Therapeutic approach
      1. Know the treatment of nasal polyps in patients with cystic fibrosis
      2. Know the treatments for distal intestinal obstruction syndrome in patients with cystic fibrosis
      3. Know the treatments for massive pulmonary hemorrhage in patients with cystic fibrosis
      4. Know the treatment options for pneumothorax in patients with cystic fibrosis
      5. Know the positive and negative effects of bronchodilators on pulmonary function in patients with cystic fibrosis
      6. Know that altered pharmacokinetics occur in patients with cystic fibrosis
      7. Know the therapeutic approach for the patient with cystic fibrosis who is exposed to varicella
      8. Know the association between pancreatic enzyme replacement therapy and colonic strictures
      9. Know the treatment of acute pulmonary exacerbation in cystic fibrosis
      10. Know the indications for the use of DNase in cystic fibrosis
      11. Know the approach to treatment of initial colonization with Pseudomonas aeruginosa in an infant with cystic fibrosis
      12. Know the indications for the use of inhaled tobramycin in cystic fibrosis
      13. Know the indications for the use of inhaled hypertonic saline in cystic fibrosis
6. Prognosis
   a. Natural history
   b. Prognosis with therapy
      1. Know the prognosis of cystic fibrosis with current therapy

F. Interstitial lung disease
   1. Epidemiology
   2. Etiology/genetics
      a. Know that bronchiolitis obliterans can follow a variety of insults, including pulmonary infections, inhalation of toxic gases, and graft-versus-host disease
      b. Know which disease entities are associated with lymphoid interstitial pneumonia
      c. Know which medications cause interstitial lung disease
      d. Know that radiation exposure can cause interstitial fibrosis
      e. Know which collagen vascular diseases produce interstitial lung disease

f. Recognize that interstitial lung disease may be a manifestation of graft versus host disease following bone marrow transplantation
g. Know the most common causes of hypersensitivity pneumonitis
h. Recognize which inborn errors of surfactant metabolism (mutations resulting in surfactant dysfunction) cause interstitial lung disease in neonates, infants, and children
i. Know that neuroendocrine cell hyperplasia of infancy (NEHI) and pulmonary interstitial glycogenosis are forms of chronic lung disease that are unique to infants
j. Know that lung growth disorders characterized by alveolar simplification are often misdiagnosed as interstitial lung disease and are the most common finding in lung biopsies done in children under 2 years of age

3. Pathophysiology
   a. Pathology
      1. Recognize the histologic picture of bronchiolitis obliterans
      2. Know the histology of desquamative interstitial pneumonitis
      3. Know the histology of lymphoid interstitial pneumonia
      4. Recognize the pulmonary manifestations of collagen vascular disease
   b. Path mechanisms and consequences
      1. Know that surfactant proteins B and C and ABCA3 gene mutations can have various pathologies, including pulmonary alveolar proteinosis, desquamative interstitial pneumonia, chronic pneumonitis of infancy, nonspecific interstitial pneumonia, and usual inter
4. Diagnosis and clinical manifestations
   a. History
      1. Know the time course of onset of radiation-induced pneumonitis
      2. Recognize clinical symptoms suggestive of interstitial lung disease in infants and older children
      3. Recognize the value of an environmental history in the evaluation of pediatric interstitial lung disease
      4. Know the clinical presentation of acute eosinophilic pneumonia
   b. Physical examination
      1. Recognize physical findings characteristic of interstitial lung disease (eg, tachypnea and crackles) in children
   c. Imaging
      1. Recognize imaging findings typical of bronchiolitis obliterans
      2. Recognize radiographic findings typical of unilateral or localized hyperlucent lung (Swyer-James or Macleod syndrome)
      3. Recognize radiographic manifestations of interstitial lung disease
      4. Recognize radiographic manifestations of pulmonary disease secondary to collagen vascular disease
      5. Know the usefulness of high-resolution computed tomography (CT scan) in the evaluation of interstitial lung disease
      6. Know the radiologic appearance and clinical implications of honey-comb lung
   d. Pulmonary function tests
      1. Identify pulmonary function abnormalities associated with interstitial lung disease

2. Recognize the lung function abnormalities (including hypoxemia) that develop during exercise in patients with severe interstitial lung disease

e. Other investigations

f. Diagnostic criteria

1. Know that open lung biopsy is usually necessary for the accurate diagnosis of interstitial lung disease

g. Complications

1. Recognize the complications associated with interstitial lung disease in childhood
2. Recognize that pulmonary hypertension is associated with a decreased probability of survival in pediatric interstitial lung disease

5. Prevention and therapeutic approach

a. Prevention

b. Therapeutic approach

1. Understand that although anti-inflammatory drugs (eg, systemic corticosteroids) are commonly used in pediatric interstitial lung disease, there is no definitive evidence to support this practice
2. Know the role of lung transplantation in the management of pediatric interstitial lung disease

c. Side effects of therapy

1. Understand the risk/benefit relationship in using systemic corticosteroids for pediatric interstitial lung disease

6. Prognosis

a. Natural history

1. Know the natural history of bronchiolitis obliterans in an immunocompetent host
2. Know the natural history of chronic hypersensitivity pneumonia in childhood
3. Know the natural history of interstitial pulmonary fibrosis in childhood
4. Know the natural history of known inborn errors of surfactant metabolism, neuroendocrine cell hyperplasia of infancy, and pulmonary interstitial glycogenosis

b. Prognosis with therapy

1. Know that the bronchiolitis obliterans associated with graft-versus-host disease is often fatal despite therapy

G. Pneumonia

1. Bacterial pneumonia

a. Epidemiology

1. Know that the incidence of acute pneumonia is greatest in children younger than 5 years of age
2. Know how the incidence of Mycoplasma pneumonia varies with age

b. Etiology/genetics

1. Know the most common pathogens responsible for bacterial pneumonia in infants under 1 month of age
2. Know the most common pathogens responsible for bacterial pneumonia in children between 1 month and 2 years of age

3. Know the most common bacteria causing pneumonia in children between 5 and 15 years of age
4. Know the most common pathogens responsible for lung abscesses in children
5. Know the most common pathogens responsible for empyema in children
6. Know the most common pathogens responsible for anaerobic bacterial pneumonia in children
7. Know the most common pathogens responsible for bacterial pneumonia in children with splenic dysfunction or absence of the spleen
8. Know the most common pathogens responsible for bacterial pneumonia in children with immunoglobulin deficiency
9. Know the most common pathogens responsible for bacterial pneumonia in pharmacologically immunosuppressed children
10. Know the common pathogens causing pneumonia in children with chronic granulomatous disease
11. Know the common pathogens causing pneumonia in children with sickle cell disease
12. Know the most common bacteria causing pneumonia in children between 4 months and 4 years of age

c. Pathophysiology
   1. Pathology
      a. Know the histologic appearance of common childhood bacterial pneumonia
      b. Know the histologic appearance of a sputum sample in acute bacterial pneumonia
   2. Path mechanisms and consequences
      a. Know the differences in inflammatory cell infiltrates in bacterial and viral pneumonia
      b. Understand the etiology of air-fluid levels in the lungs in patients with bacterial pneumonia

d. Diagnosis and clinical manifestations
   1. History
      a. Recognize the symptoms of acute pneumonia in infants
      b. Recognize the symptoms of acute pneumonia in children
      c. Know the risk factors for anaerobic pneumonia in children
      d. Know the findings in the history that suggest Mycoplasma pneumonia
   2. Physical examination
      a. Recognize the physical findings associated with acute lobar pneumonia in infants
      b. Recognize the physical findings associated with acute lobar pneumonia in older children
      c. Know that malodorous breath may signify anaerobic bacterial pneumonia
      d. Recognize discrepancies between radiographic and physical findings in children infected with Mycoplasma pneumonia
      e. Know the typical physical examination findings in pleural effusion
      f. Know the typical physical examination findings in pulmonary consolidation
   3. Imaging
      a. Recognize the radiographic appearance of pneumatoceles

b. Know that radiographic signs do not distinguish between viral and bacterial pneumonias
c. Know the typical appearance of a lobar pneumonia on imaging studies of the chest
d. Know the typical appearance of a lung abscess on imaging studies of the chest
e. Know the typical appearance of a pleural effusion on a radiographic study of the chest
f. Know the appearance of pleural fluid and/or empyemas on an ultrasound study of the chest
g. Know that cylindrical bronchiectasis is a common finding during resolution of uncomplicated pneumonia in an otherwise normal child
h. Know the natural history of radiographic changes during and after pneumonia
i. Know the role of imaging procedures in obtaining diagnostic samples or performing therapeutic drainage of pleural fluid or lung abscesses

4. Pulmonary function tests
   a. Know the changes in blood gas tensions that accompany acute pneumonia
   b. Know the changes in spirometry that would be expected in acute lobar pneumonia
5. Other investigations
   a. Know the limitations of antigen tests for bacterial pneumonias in children
   b. Know the frequency of positive blood cultures associated with bacterial pneumonia in childhood is low
   c. Know the limitations of nasal and oral bacterial cultures in a child with pneumonia
   d. Know the procedure for obtaining and the limitations of bacterial cultures from bronchoscopy in the diagnosis of bacterial pneumonias
   e. Know the typical laboratory characteristics of pleural fluid in empyema
   f. Know the typical laboratory characteristics of a parapneumonic effusion complicating a pneumonia
   g. Know the indications for pleural space drainage of pleural effusion in patients with pneumonia
6. Diagnostic criteria
   a. Understand the role of bronchoalveolar lavage and lung biopsy in diagnosing pneumonia
7. Complications

e. Prevention and therapeutic approach
   1. Prevention
      a. Know the impact of Haemophilus influenzae type B vaccination on pneumonia in normal and immunocompromised patients
      b. Know the impact of 21-valent S. pneumonia vaccination on pneumonia in immunocompromised patients
      c. Know that prophylactic installation of antibiotics in the eyes at birth does not prevent the later onset of chlamydial pneumonia
   2. Therapeutic approach

a. Know which antibiotics adequately treat infection with Mycoplasma pneumoniae
b. Know appropriate antibiotic therapy for treatment of anaerobic bacterial pneumonia
c. Know appropriate empiric therapy for immunodeficient patients with pneumonia
d. Know how body position may affect oxygenation in patients with lobar pneumonia
e. Know the indications and methods for oxygen therapy in bacterial pneumonia
f. Know that inappropriate secretion of antidiuretic hormone (ISADH) may complicate severe pneumonias and how ISADH is diagnosed and treated
g. Understand that chest physiotherapy has no role in management of uncomplicated lobar pneumonia in an otherwise healthy child
h. Understand the management of lung abscess
i. Understand the management of empyema
j. Know the role of antibiotics in the treatment of Bordetella pertussis infection
k. Know the appropriate empiric therapy for community-acquired pneumonia in immunocompetent infants, children, and adolescents

3. Side effects of therapy
a. Know complications of drainage of pulmonary abscesses
b. Know the risks associated with bronchoscopy in the diagnosis and management of pulmonary abscess

f. Prognosis
1. Natural history
a. Know the long-term consequences of empyemas
2. Prognosis with therapy
a. Know the natural history of pleural thickening in a patient successfully treated for empyema
b. Know the natural history of pneumonia with and without therapy

2. Viral pneumonia
a. Epidemiology
1. Understand seasonal variation in the frequency of viral pneumonia caused by different viral pathogens

b. Etiology/genetics
1. Know the common viruses causing pneumonia in the newborn period
2. Know the common viral etiologies of pneumonia after the neonatal period
3. Know the most common pathogens responsible for viral pneumonia in pharmacologically immunosuppressed patients

c. Pathophysiology
1. Pathology
a. Know the histologic appearance of common viral pneumonias in pediatric patients
2. Path mechanisms and consequences

d. Diagnosis and clinical manifestations

1. History
2. Physical examination
    a. Recognize the similarity of physical findings in viral and bacterial pneumonia
3. Pulmonary function tests
4. Other investigations
    a. Know the utility and limitations of rapid diagnostic tests for viral pneumonia
5. Diagnostic criteria
    a. Understand the roles of serology and cultures in the diagnosis of viral pneumonias
6. Complications
    a. Know that bacterial superinfection may complicate viral pneumonia

e. Therapeutic approach and side effects (Prevention - see Section IV.A)
1. Therapeutic approach
    a. Know the indications for the use of antiviral agents in the treatment of viral pneumonias
2. Side effects of therapy
    a. Know the side effects of antiviral agents

f. Prognosis
1. Natural history
2. Prognosis with therapy
    a. Know the natural history of cytomegalovirus pneumonitis

3. Fungal pneumonia
a. Epidemiology
1. Know that fungi can be pathogens in the respiratory tract in a susceptible host
2. Know the geographic variation in pathogens causing fungal pneumonias

b. Etiology/genetics

c. Pathophysiology
1. Pathology
    a. Recognize the histologic appearance of fungal infections
    b. Know that Cryptococcus neoformans has a distinguishing polysaccharide capsule
2. Path mechanisms and consequences
    a. Know that inhalation of fungal spores causes respiratory disease with Histoplasma, Blastomyces, Coccidioides, and Cryptococcus
    b. Know that fungal infections can develop as a result of dysfunction of lymphocytes, macrophages, neutrophils and/or complement

d. Diagnosis and clinical manifestations
1. History
    a. Know that fungal pulmonary infection may cause acute or insidious onset of symptoms or may be asymptomatic
2. Physical examination
3. Imaging
4. Pulmonary function tests
5. Other investigations

a. Know the role of antigen detection tests in the diagnosis of fungal infections of the lung

6. Diagnostic criteria
   a. Know that the clinical and radiographic picture of pulmonary histoplasmosis may be similar to that of pulmonary tuberculosis
   b. Know that the diagnosis of fungal infections can be confirmed by measurement of titers during the acute and convalescent phases
   c. Know the radiographic appearance of fungal pulmonary infections
7. Complications
   a. Know that extrapulmonary disease can occur in patients with fungal pneumonias

e. Prevention and therapeutic approach
   1. Prevention
   2. Therapeutic approach
      a. Know the treatment for pulmonary fungal infections
   3. Side effects of therapy
      a. Know the side effects of drugs commonly prescribed for pulmonary fungal infections

f. Prognosis
   1. Natural history
      a. Know the natural history of common pulmonary fungal infections in immunocompetent and immunocompromised hosts
   2. Prognosis with therapy

4. Tubercle bacilli
   a. Epidemiology
      1. Know that miliary tuberculosis and lymphatic tuberculosis occur more frequently in patients younger than 4 years of age
      2. Know the epidemiology of drug-resistant tuberculosis
      3. Know the epidemiology of atypical mycobacteria
   b. Etiology/genetics
      1. Know the factors that increase risk for primary tuberculosis in children
      2. Know the circumstances that predispose to reactivation of tuberculosis
      3. Know why children with active tuberculosis rarely infect others
   c. Pathophysiology
      1. Pathology
         a. Recognize the typical histologic appearance of a caseating granuloma
         b. Know that atypical mycobacteria are more likely to cause mycobacterial cervical adenitis than is Mycobacterium tuberculosis
         c. Recognize the typical histologic appearance of acid-fast bacilli
         d. Understand that transplacental infection of the fetus with Mycobacterium tuberculosis may occur
      2. Path mechanisms and consequences
         a. Know the mechanism of immune response to infection with Mycobacterium tuberculosis
         b. Understand that mycobacteria in primary infection are cleared via lymphatics

d. Diagnosis and clinical manifestations
    1. History
        a. Know that the risk for the development of tuberculous disease is greatest within the first two years after initial exposure
        b. Know that tuberculosis meningitis may have an insidious onset with a prolonged nonspecific prodrome of gastrointestinal symptoms
    2. Physical examination
        a. Know that physical examination of children with active tuberculosis may reveal normal findings or non-specific signs
    3. Imaging
        a. Recognize miliary tuberculosis on an x-ray study of the chest
        b. Recognize the typical findings of primary tuberculosis on an x-ray study of the chest
    4. Pulmonary function tests
    5. Other investigations
        a. Know the technique and diagnostic yield of gastric washing for Mycobacterium tuberculosis culture
        b. Know the influence of bacille Calmette-Guerin immunizations on tuberculin skin testing
        c. Know the etiologies of false-negative results of intradermal tuberculin skin tests
        d. Identify the etiology of false-positive results of intradermal tuberculin skin tests
        e. Know the utility of bronchoalveolar lavage in the diagnosis of pulmonary tuberculosis
        f. Know the influence of previous or current infection with atypical mycobacteria on tuberculin skin testing
        g. Know how to interpret results of a tuberculin skin test in a patient with immunodeficiency
        h. Know how to interpret results of a tuberculin skin test in immunocompetent patients at low versus high risk of tuberculosis exposure
    6. Diagnostic criteria
        a. Know the likelihood of culturing Mycobacterium tuberculosis from gastric aspirates in various clinical stages of tuberculosis (eg, primary versus cavitary)
        b. Know how to evaluate a patient for tuberculosis, given that BCG vaccine was previously administered
    7. Complications
        a. Understand the pulmonary complications of untreated primary tuberculosis
e. Prevention and therapeutic approach
    1. Prevention
        a. Know the management of a newborn infant whose mother has active tuberculosis
        b. Describe appropriate methods to evaluate family members in the home of a child with positive results on a tuberculin skin test

c. Understand the role of community surveillance in the management of tuberculosis

2. Therapeutic approach
   a. Know the recommended courses of therapy for pulmonary tuberculosis
   b. Know the toxicities of drugs commonly used for tuberculosis (i.e., isoniazid, rifampin, streptomycin, pyrazinamide, and ethambutol)
   c. Know the standard drugs and duration of treatment for acute miliary tuberculosis
   d. Know the standard drugs and the duration of treatment for tuberculous meningitis
   e. Know the standard drugs and the duration of treatment for reactivation tuberculosis
   f. Understand the rationale for directly observed therapy for pulmonary tuberculosis
3. Side effects of therapy
   a. Know the contraindications for isoniazid therapy
   b. Understand the treatment implications of multi-drug resistant pulmonary tuberculosis
   c. Understand the rationale for pyridoxine therapy during isoniazid therapy

f. Prognosis
1. Natural history
   a. Know that most primary tuberculosis infections will improve without treatment
2. Natural history with therapy
   a. Understand the effect treatment of primary tuberculosis has on the incidence of subsequent reactivation of the disease

5. Parasites
a. Epidemiology
   1. Recognize the association of pneumocystis pneumonia immunodeficiency
b. Etiology/genetics
   1. Know the routes whereby parasitic infections are acquired
c. Pathophysiology
   1. Pathology
      a. Know the various presenting characteristics of infection with Pneumocystis jiroveci (carinii)
      b. Know that pneumonitis due to Pneumocystis jiroveci (carinii) is characterized by an intra-alveolar foamy eosinophilic exudate
      c. Know that reduced or abnormal T-lymphocytes predispose to Pneumocystis jiroveci (carinii) infection
   2. Path mechanisms and consequences
d. Diagnosis and clinical manifestations
   1. History
      a. Recognize clinical manifestations of visceral larva migrans
      b. Recognize the clinical presentation of pneumocystis pneumonia
      c. Recognize clinical features of echinococcal infection

d. Know that hemoptysis may be a primary manifestation of pulmonary infection with Echinococcus

2. Physical examination
    a. Recognize the findings on physical examination of a patient with Pneumocystis jiroveci (carinii) pneumonia
    b. Recognize that clubbing may occur in patients with pulmonary parasitic infections
3. Imaging
    a. Recognize the typical appearance of Pneumocystis jiroveci (carinii) pneumonia on radiographic study of the chest
    b. Recognize radiographic features of echinococcal infection
4. Pulmonary function tests
    a. Know that restrictive lung disease on pulmonary function testing can occur with pneumonitis due to Pneumocystis jiroveci (carinii)
5. Other investigations
    a. Know the appearance of bronchoalveolar lavage fluid in pneumocystis pneumonia
    b. Know that visceral larval migrans is associated with marked eosinophilia within peripheral blood
6. Diagnostic criteria
    a. Know laboratory diagnostic procedures for pneumocystis as etiology of pneumonia
7. Complications

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
    a. Know the treatment for pneumonitis due to Pneumocystis jiroveci (carinii)
    b. Know the indications and approaches for prophylaxis against Pneumocystis jiroveci (carinii) pneumonia
3. Side effects of therapy

f. Prognosis
1. Natural history
2. Prognosis with therapy
    a. Know that survivors of Pneumocystis jiroveci (carinii) pneumonia can have normal pulmonary function test results

6. Others
a. Epidemiology
b. Etiology/genetics
c. Pathophysiology
1. Pathology
2. Path mechanisms and consequences

d. Diagnosis and clinical manifestations
1. History
2. Physical examination
3. Imaging

a. Know which pneumonia-causing organisms are not associated with pleural effusions
4. Pulmonary function tests
5. Other investigations
6. Diagnostic criteria
a. Know that chlamydial pneumonia is accompanied by eosinophilia
7. Complications
e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
3. Side effects of therapy
f. Prognosis
1. Natural history
2. Prognosis with therapy

H. Disorders of the chest wall, diaphragm, and pleural space
1. Chest wall
a. Ribs
1. Etiology/genetics
a. Know that asphyxiating thoracic dystrophy is inherited in an autosomal recessive manner
b. Recognize the inheritance and genetic mechanism underlying asphyxiating thoracic dystrophy
c. Know that abnormalities of the ventral abdominal wall, eg, giant omphalocele, can result in a long thin chest wall and pulmonary hypoplasia
2. Pathophysiology
a. Pathology
b. Path mechanisms and consequences
1. Know that the chest wall abnormalities associated with asphyxiating thoracic dystrophy (short horizontal ribs and flared costochondral junctions) result in a fixed chest wall and hypoplastic lungs
2. Know the pathophysiologic consequences of chest wall restriction
3. Diagnosis and clinical manifestations
a. History
b. Physical examination
1. Recognize the association between a small, bell-shaped chest and chronically reduced rib excursion
c. Imaging
1. Recognize the radiographic appearance of the rib cage characteristic of asphyxiating thoracic dystrophy
2. Recognize the radiographic appearance of abnormal thoracic vertebrae and ribs
3. Recognize the radiographic appearance of the chest characteristic of progressive spinal atrophy
4. Recognize the radiographic appearance of benign chest wall tumors
d. Pulmonary function tests

1. Recognize that asphyxiating thoracic dystrophy is associated with severe restrictive lung disease
2. Recognize the pulmonary function abnormalities associated with muscular dystrophy

e. Other investigations
f. Diagnostic criteria
g. Complications
1. Recognize that respiratory failure is the primary cause of death in asphyxiating thoracic dystrophy
2. Recognize that asphyxiating thoracic dystrophy is associated with polydactyly and renal and hepatic disorders

4. Prevention and therapeutic approach
a. Prevention
b. Therapeutic approach
1. Know that surgical approaches like lateral chest expansion have been used with varying degrees of success in asphyxiating thoracic dystrophy

5. Prognosis
a. Natural history
1. Recognize that the course of asphyxiating thoracic dystrophy is variable and is dependent on the degree of pulmonary hypoplasia

b. Prognosis with therapy

b. Sternum-pectus deformities
1. Epidemiology
2. Etiology/genetics
a. Recognize that pectus excavatum occurs as both a congenital and acquired defect, the latter most likely secondary to respiratory disease with increased work of breathing
b. Recognize that pectus excavatum is associated with Marfan syndrome

3. Pathophysiology
a. Pathology
b. Path mechanisms and consequences

4. Diagnosis and clinical manifestations
a. History
b. Physical examination
c. Imaging
1. Recognize findings associated with a pectus excavatum deformity on x-ray study of the chest

d. Pulmonary function tests
1. Recognize changes in lung function secondary to a pectus excavatum deformity
2. Recognize the effects of pectus excavatum on exercise tolerance
3. Realize that pectus carinatum does not cause significant changes in cardiopulmonary function

e. Other investigations
f. Diagnostic criteria
g. Complications

1. Recognize that asymmetric pectus excavatum may be associated with secondary thoracic scoliosis

5. Prevention and therapeutic approach
   a. Prevention
   b. Therapeutic approach
      1. Recognize that the primary indication for surgical treatment of pectus deformities is cosmesis
      2. Realize that surgical correction of pectus excavatum deformity is not associated with significant improvement in pulmonary function
      3. Recognize that in a small number of patients with pectus excavatum, exercise tolerance is limited by the inability to increase tidal volume at maximal exercise workload
      4. Know that moderate pectus excavatum does not cause symptoms at rest or on exercise
   c. Side effects of therapy
      1. Know that too early or too extensive a repair of pectus excavatum can result in failure of chest wall growth and development of acquired asphyxiating thoracic dystrophy
6. Prognosis
   a. Natural History
      1. Recognize that a pectus excavatum deformity may worsen with growth
   b. Prognosis with therapy
      1. Recognize that recurrence of a pectus excavatum is possible following surgical repair, especially in children with Marfan syndrome

c. Diaphragm
   1. Epidemiology
   2. Etiology/genetics
      a. Recognize birth factors and/or surgical procedures that may lead to diaphragmatic paralysis
   3. Pathophysiology
      a. Pathology
      b. Path mechanisms and consequences
         1. Know that eventration of the diaphragm can result from diaphragmatic maldevelopment
         2. Know that unilateral diaphragmatic weakness is most problematic in early infancy due to the nature of the rib cage (position and compliance)
   4. Diagnosis and clinical manifestations
      a. History
         1. Recognize that diaphragmatic eventration is included in the differential diagnosis of severe respiratory distress in newborn infants
         2. Recognize that diaphragmatic eventration can be associated with breech or face presentation or difficult forceps extraction during delivery
      b. Physical examination
         1. Recognize the physical findings associated with diaphragmatic eventration

2. Recognize the physical findings typical of unilateral and bilateral diaphragmatic paralysis

c. Imaging
1. Recognize the radiographic appearance of diaphragmatic eventration
2. Recognize the radiographic appearance of diaphragmatic paralysis
3. Recognize the significance of paradoxical movement of the diaphragm on fluoroscopy

d. Pulmonary function tests
e. Other investigations
f. Diagnostic criteria
g. Complications
1. Recognize that chronic atelectasis and chronic/recurrent pneumonia are complications of diaphragmatic eventration and paralysis

5. Prevention and therapeutic approach
a. Prevention
b. Therapeutic approach
1. Recognize the indications for repair of a diaphragmatic eventration
2. Know that conservative management using an endotracheal tube and continuous positive airway pressure is indicated in patients with postsurgical traumatic injury of the phrenic nerve
3. Know that persistence of symptoms that require continued ventilatory support following post-surgical traumatic injury of the phrenic nerve is an indication for plication of the diaphragm
4. Know that the best position for an infant with unilateral diaphragmatic paralysis is with the affected diaphragm down

c. Side effects of therapy

6. Prognosis
a. Natural history
1. Know the natural history of postsurgical traumatic injury of the phrenic nerve

b. Prognosis with therapy

d. Thoracic vertebrae - scoliosis
1. Epidemiology
2. Etiology/genetics
a. Recognize that the majority of cases of scoliosis are idiopathic

3. Pathophysiology
a. Pathology
b. Path mechanisms and consequences

4. Diagnosis and clinical manifestations
a. History
b. Physical
c. Imaging
1. Recognize changes induced by thoracic scoliosis on x-ray study of the chest

d. Pulmonary function tests

1. Recognize that curvatures greater than 60 degrees are associated with detectable pulmonary function abnormalities
2. Recognize that scoliosis may result in restrictive or obstructive or mixed pulmonary function abnormalities
3. Recognize that residual volume is normal and total lung capacity is decreased in patients with thoracic scoliosis
4. Recognize that VD/VT increases with severe scoliosis, contributing to alveolar hypoventilation
5. Recognize that chest wall compliance is reduced more than lung compliance in patients with scoliosis
6. Recognize that exercise tolerance in patients with thoracic scoliosis is usually limited by ventilatory rather than circulatory factors

e. Other investigations
f. Diagnostic criteria
g. Complications
   1. Recognize the relationship between curvatures greater than 90 degrees and the increased risk of cardiorespiratory failure in patients with scoliosis
   2. Recognize that hypoventilation and hypoxemia may occur during REM sleep in patients with moderate scoliosis
   3. Recognize that pulmonary hypertension is a complication of severe scoliosis
   4. Know the association between thoracic vertebral rotation and central airway obstruction

5. Prevention and therapeutic approach
   a. Prevention
   b. Therapeutic approach
      1. Know that surgical intervention in children and adolescents with scoliosis may be indicated to prevent worsening of lung function
   c. Side effects of therapy
      1. Recognize that atelectasis, hemothorax, pneumothorax, pulmonary edema, and fat emboli are complications of surgical management of scoliosis
      2. Recognize that underlying neuromuscular disease increases the risks of postoperative complications in patients with scoliosis
      3. Recognize that scoliosis surgery in patients with muscular weakness (SMA, Duchenne muscular dystrophy) does not improve lung function or prevent progression of pulmonary complications and respiratory failure
6. Prognosis
   a. Natural history
   b. Prognosis with therapy

2. Pleural space
   a. Effusion
      1. Epidemiology
      2. Etiology/genetics

a. Know the etiologies of chylothorax
b. Know that an imbalance between fluid reabsorption and filtration determines the accumulation of fluid in the pleural space
c. Know the subdiaphragmatic processes that may lead to pleural effusions
d. Know the etiologies of pleural effusion

3. Pathophysiology
   a. Pathology
   b. Path mechanisms and consequences
      1. Know the pathophysiology of parapneumonic effusions
      2. Know that lymphoma may be associated with pleural effusions
      3. Know that congenital pulmonary lymphangiectasia is associated with chylous effusion in the neonatal period
4. Diagnosis and clinical manifestations
   a. History
   b. Physical examination
      1. Recognize the physical findings characteristic of a pleural effusion
   c. Imaging
      1. Recognize the limitations of a supine-view x-ray study of the chest in detecting pleural effusion
      2. Recognize findings typical of a pleural effusion on x-ray study of the chest
      3. Recognize the role of lateral/decubitus-view x-ray studies of the chest in the evaluation of a pleural effusion
      4. Recognize the role of ultrasonography and computed tomography (CT scan) of the chest in the evaluation of a pleural effusion
   d. Pulmonary function tests
   e. Other investigations
      1. Recognize the physical and chemical characteristics typical of pleural effusions due to various causes (eg, chylous effusion, simple transudate, exudate)
      2. Recognize the indications for thoracentesis in a child with a pleural effusion
      3. Know the proper procedure for performing thoracentesis in a child
      4. Know the differential diagnosis of a bloody pleural effusion
   f. Diagnostic criteria
   g. Complications
5. Prevention and therapeutic approach
   a. Prevention
   b. Therapeutic approach
      1. Know the management of malignant pleural effusions
      2. Know the management of chylothorax
      3. Know the management of pleural effusions (transudates and exudates)
      4. Know the management of empyema
   c. Side effects of therapy
6. Prognosis
   a. Natural history

b. Prognosis with therapy
    1. Know the natural history of treated empyema

b. Pneumothorax
1. Epidemiology
    a. Recognize conditions associated with increased risk of spontaneous pneumothorax
2. Etiology/genetics
3. Pathophysiology
    a. Pathology
    b. Path mechanisms and consequences
4. Diagnosis and clinical manifestations
    a. History
        1. Know that spontaneous pneumothorax is a common cause of chest pain in adolescents
    b. Physical examination
        1. Recognize the physical findings associated with a pneumothorax (shift of trachea and cardiac apex, and a tympanic percussion note over the affected side)
        2. Recognize the significance of a bulging hemithorax in a neonate
    c. Imaging
        1. Recognize radiographic findings typical of pneumothorax
        2. Know how to distinguish lobar emphysema and a pulmonary cyst from a pneumothorax
    d. Pulmonary function tests
    e. Other investigations
    f. Diagnostic criteria
    g. Complications
5. Prevention and therapeutic approach
    a. Prevention
    b. Therapeutic approach
        1. Know the appropriate management of a spontaneous pneumothorax
        2. Know the appropriate management of recurrent pneumothorax in a normal child
        3. Know the appropriate management of recurrent pneumothorax in a child with cystic fibrosis
        4. Know the physiologic principles underlying the use of 100% oxygen in the treatment of pneumothorax
        5. Know the methods for pleurodesis
        6. Know the risks of air transport in patients with air leak syndromes
    c. Side effects of therapy
        1. Know that infection, pain, bleeding, and respiratory splinting are complications of chest tube drainage
        2. Know that loculated pneumothoraces may be difficult to drain with a single chest tube
        3. Recognize complications of chemical pleurodesis
6. Prognosis

a. Natural history
    1. Recognize the risk of recurrence of a pneumothorax in children with obstructive lung disease (ie, cystic fibrosis)
b. Prognosis with therapy
    1. Know that pleural abrasion is more effective than chemical agents in reducing the risk of recurrent pneumothorax in patients with cystic fibrosis

c. Mediastinum
1. Epidemiology
2. Etiology/genetics
    a. Know that histoplasmosis is the most common cause of fibrosing mediastinitis
3. Pathophysiology
    a. Pathology
    b. Path mechanisms and consequences
        1. Know the pathophysiology of pneumomediastinum
4. Diagnosis and clinical manifestations
    a. History
        1. Recognize the clinical manifestations of a mediastinal mass
        2. Recognize the clinical picture of acute bacterial mediastinitis
    b. Physical examination
        1. Know that mediastinal masses may not be apparent on physical examination
        2. Know that the respiratory findings typical of mediastinal masses are generally secondary to airway compression
        3. Recognize the importance of evaluating the patient with a mediastinal mass for superior vena cava syndrome
        4. Recognize the importance of evaluating pulsus paradoxus in the patient at risk for cardiac tamponade or venous obstruction in a patient with a large mediastinal mass
    c. Imaging
        1. Know the differential diagnosis of the radiographic finding of a mass in each of the four compartments of the mediastinum
        2. Recognize indications for plain x-ray studies of the chest in the evaluation of a mediastinal mass
        3. Recognize indications for computed tomography (CT scan) of the chest in the evaluation of a mediastinal mass
        4. Recognize indications for magnetic resonance imaging (MRI) in the evaluation of a mediastinal mass
        5. Recognize the radiographic appearance of a pneumomediastinum
        6. Know which masses are likely to occur in the anterior, posterior, middle, and superior mediastinum
    d. Pulmonary function tests
        1. Know that a patient with a mediastinal mass may have a pulmonary function test pattern suggestive of a fixed intrathoracic obstruction
    e. Other investigations

1. Recognize the risks associated with bronchoscopy and/or general anesthesia in a child with tracheal compression due to a mediastinal mass
   f. Diagnostic criteria
   g. Complications
5. Prevention and therapeutic approach
   a. Prevention
   b. Therapeutic approach
      1. Recognize that pneumomediastinum in an older child rarely requires therapy
   c. Side effects of therapy
6. Prognosis
   a. Natural history
   b. Prognosis with therapy

I. Acute lung diseases in the newborn infant
1. Epidemiology
2. Etiology/genetics
   a. Know that spinal muscular atrophy type I is an inherited disorder
   b. Know the etiology of transient tachypnea of the newborn
   c. Know the etiology of meconium aspiration syndrome
   d. Know the etiology of neonatal respiratory distress syndrome
   e. Know the most common causes of congenital pneumonia in neonates
   f. Know the etiology of persistent pulmonary hypertension of the neonate
3. Pathophysiology
   a. Pathology
      1. Recognize the pathology of neonatal respiratory distress syndrome on a histopathologic section
   b. Path mechanisms and consequences
      1. Know the pathophysiologic consequences of high surface tension at the air liquid interface (eg, bronchiolar epithelial injury, pulmonary edema, low end expiratory volume, etc)
      2. Know the mechanisms contributing to hypoxemia in respiratory distress syndrome
      3. Know the mechanisms of meconium aspiration lung injury
      4. Understand how surfactant deficiency in neonatal respiratory distress syndrome affects the relative surface tension of small versus large alveoli in terms of LaPlace's law
      5. Know the mechanisms contributing to hypoxemia in meconium aspiration syndrome
      6. Know the mechanisms contributing to persistent pulmonary hypertension of the neonate
      7. Know the mechanisms contributing to hypoxemia in persistent pulmonary hypertension of the neonate
4. Diagnosis and clinical manifestations
   a. History
      1. Recognize elements of the maternal history that increase or decrease the likelihood of neonatal respiratory distress syndrome

2. Recognize elements of the maternal history that increase the likelihood of neonatal pneumonia
3. Recognize elements of the maternal history that increase the likelihood of meconium aspiration syndrome
4. Recognize elements of the maternal history that increase the likelihood of persistent pulmonary hypertension

b. Physical examination
1. Recognize the expected values of preductal versus postductal pulse oximetry readings in a neonate with persistent pulmonary hypertension

c. Imaging
1. Recognize the radiographic features of meconium aspiration lung injury
2. Recognize the radiographic presentation of transient tachypnea of the newborn
3. Recognize the radiographic features of neonatal respiratory distress syndrome

d. Pulmonary function tests
1. Know that specific airway resistance is normal in neonatal respiratory distress syndrome
2. Know that lung compliance is reduced in neonatal respiratory distress syndrome

e. Other investigations

f. Diagnostic criteria
1. Know the differential diagnosis of airflow obstruction in newborn infants
2. Know that early onset streptococcal pneumonia may mimic respiratory distress syndrome
3. Know the diagnostic criteria for persistent pulmonary hypertension of the neonate

g. Complications
1. Recognize complications of treatment of neonatal respiratory distress syndrome
2. Recognize complications of severe meconium aspiration
3. Recognize the complications of persistent pulmonary hypertension of the neonate and its treatment

5. Prevention and therapeutic approach

a. Prevention
1. Know that prophylactic surfactant therapy may prevent or lessen the severity of neonatal respiratory distress syndrome
2. Know that antenatal corticosteroids are used to prevent or lessen the severity of neonatal respiratory distress syndrome

b. Therapeutic approach
1. Know that neonatal respiratory distress syndrome may be treated with surfactant replacement therapy
2. Know that persistent pulmonary hypertension of the neonate may be treated with inhaled nitric oxide
3. Know that primary pulmonary hypertension of the neonate unresponsive to conventional therapy may be treated with extracorporeal membrane oxygenation (ECMO)

c. Side effects of therapy
1. Know the complications of therapy for neonatal respiratory distress syndrome
2. Know the effect of postnatal corticosteroid therapy on neurodevelopment

6. Prognosis
    a. Natural history
    b. Prognosis with therapy
        1. Know that long-term sequelae (childhood and adult life) of neonatal lung injury (RDS, meconium aspiration, extremely low birth weight, etc.) include airway obstruction, hyperinflation, and airway hyperreactivity
        2. Know that some infants with neonatal lung injury develop chronic lung disease of infancy (bronchopulmonary dysplasia)

J. Respiratory failure
1. Epidemiology
    a. Know that death due to respiratory failure is greater in the first year after birth than in any other year of childhood
2. Etiology/genetics
    a. Recognize the extra-pulmonary causes of respiratory failure (eg, central nervous system injury, severe chest wall deformity, or ventilatory muscle dysfunction)
    b. Know that pulmonary causes of respiratory failure are due to excessively increased work of breathing and/or severe gas exchange abnormalities
3. Pathophysiology
    a. Pathology
    b. Path mechanisms and consequences
        1. Know the metabolic compensation for chronic respiratory failure
        2. Know the compensatory mechanisms that preserve oxygen delivery to tissues in the presence of chronic hypoxemia
        3. Know the consequences of anemia upon oxygen delivery to tissues at rest and during exercise
        4. Know the formula for calculating tissue oxygen delivery and its clinical application
        5. Know the clinical factors affecting oxygen delivery
        6. Recognize the clinical signs of inadequate oxygen delivery
        7. Know the clinical consequences of acute hypoxemia
        8. Know the clinical consequences of chronic hypoxemia
        9. Know the consequences of acute respiratory acidosis
        10. Distinguish the physiologic consequences of acute hypercapnia from those of chronic hypercapnia
        11. Recognize that hypoventilation may occur during sleep but be absent when awake
        12. Know that decreased strength and/or endurance of the diaphragm can cause inspiratory muscle fatigue and respiratory failure
        13. Recognize how failure of the respiratory pump affects gas exchange
        14. Know that hyperinflation of the lungs puts the diaphragm at a mechanical disadvantage and contributes to respiratory failure
        15. Know that hypoxia, hypercapnia, and malnutrition decrease endurance of the diaphragm by decreasing muscle cell energy production, contributing to respiratory failure

16. Know that strength and endurance of the infant's diaphragm is less than that of pubertal children and adults, potentially contributing to diaphragm fatigue and respiratory failure
17. Know that increased respiratory loads contribute to diaphragm fatigue and respiratory failure
18. Know that decreased or absent central respiratory drive can cause respiratory failure and that this will be worse in the presence of increased respiratory loads

4. Diagnosis and clinical manifestations
   a. History
      1. Recognize the symptoms of recurrent nocturnal hypoxemia and hypercapnia
      2. Recognize that frequent and/or severe pneumonia in children with neuromuscular disease indicates ventilatory muscle weakness and an increased likelihood of respiratory failure
      3. Recognize that children with progressive neuromuscular diseases and disorders of respiratory control manifest symptoms first during sleep and only later during wakefulness
      4. Recognize that morning headaches may indicate hypercapnia and/or hypoxemia occurring during sleep
      5. Recognize that respiratory failure may appear to occur suddenly in children with chronic disorders of respiratory control or ventilatory muscle function, even though they have a chronic disorder affecting respiration
   b. Physical examination
      1. Understand that patients with severe neuromuscular disease and chronic respiratory failure may not manifest clinical symptoms, such as retractions and nasal flaring
      2. Recognize the physical findings typical of acute hypoxemia
      3. Recognize the physical findings typical of acute carbon dioxide retention
      4. Recognize the signs of respiratory failure in patients with severe neuromuscular weakness (e.g., tachycardia, diaphoresis)
      5. Understand that symptoms normally associated with respiratory distress (tachypnea, retractions, nasal flaring, etc.) require intact ventilatory control
      6. Recognize that children with disorders of respiratory control may not manifest respiratory distress or typical symptoms (retractions, nasal flaring, tachypnea, etc.)
   c. Imaging
      1. Recognize the radiologic findings in spinal muscular atrophy and other neuromuscular diseases
      2. Know how to use fluoroscopy and/or abdominal ultrasound to diagnose unilateral and bilateral diaphragm paralysis
   d. Pulmonary function tests
      1. Recognize that spirometry and blood gas tensions correlate poorly in patients with respiratory failure
   e. Other investigations
      1. Know the age-dependent normal values for $PaO_2$, $PaCO_2$, and pH
      2. Know the techniques for and complications of obtaining arterial blood samples
      3. Recognize the effects of sample errors on blood gas values

4. Recognize alveolar hypoventilation and hypoxemia from blood gas data
5. Recognize metabolic and respiratory acidosis from blood gas data
6. Know the effect of sampling sites on arterial blood gas values in patients with patent ductus arteriosus and right-to-left shunts
7. Recognize changes in arterial blood gas values secondary to hyperventilation due to anxiety
8. Recognize the measurement errors associated with capillary blood gas and venous blood gas tension analysis
9. Recognize various laboratory findings for different causes of hypoxemia
10. Know conditions under which measurements of end-tidal CO2 will correlate poorly with arterial PCO2 (i.e., severe obstructive lung disease)

f. Diagnostic criteria
1. Know that chronic respiratory failure occurs in a patient who cannot be weaned after at least 1 month of consistent attempts while medically stable or with an irreversible diagnosis causing respiratory failure (e.g., high spinal cord injury or CCHS)

5. Prevention and therapeutic approach
a. Prevention
1. Know which drugs produce central respiratory depression and the treatment for such depression
2. Know which drug interactions predispose patients to respiratory center depression and respiratory failure
3. Know that respiratory failure can be anticipated in children with progressive neuromuscular diseases and that discussions about long- term treatment should be held with families before this occurs
4. Recognize that impairment of airway clearance can occur before respiratory failure in patients with neuromuscular disease
5. Know that impaired airway clearance predisposes towards respiratory failure in patients with neuromuscular weakness

b. Therapeutic approach
1. Know the effect of blood transfusion and calculate its impact on oxygen delivery and oxygen content
2. Know the indications for mechanical ventilation in patients with respiratory failure
3. Understand how hyperbaric oxygen therapy might improve oxygen transport to tissues
4. Know how to treat respiratory alkalosis
5. Know the treatment of altitude-induced pulmonary edema
6. Know the oxygen-carrying capacity of hemoglobin
7. Know the differences in goals of mechanical assisted ventilation in children with acute versus chronic respiratory failure

c. Side effects of therapy
1. Recognize that mechanical ventilation in the treatment of chronic respiratory acidosis resulting in a rapid fall in PCO2 may lead to alkalosis and seizures

6. Prognosis
a. Natural history

b. Prognosis with therapy

K. Aspiration/inhalation injuries
1. Foreign body aspiration
a. Epidemiology
1. Know that the majority of patients who aspirate foreign bodies are 3 years old or less
b. Etiology/genetics
1. Recognize retained foreign body aspiration as an etiology of recurrent pneumonia
2. Know that in patients with symptomatic foreign body aspiration 80% are the result of peanut aspiration
3. Know that many cases of aspiration are not observed, resulting in delayed diagnosis
c. Pathophysiology
1. Pathology
2. Path mechanisms and consequences
a. Describe why the lung distal to a foreign body can be either overinflated or underinflated
b. Recognize the roles of esophageal foreign body in producing airway obstruction
c. Understand the mechanisms of hemoptysis following foreign body aspiration
d. Know the mechanisms by which a retained foreign body leads to bronchiectasis
e. Understand the acute pathophysiologic consequences of foreign body aspiration in the mainstem bronchus
d. Diagnosis and clinical manifestations
1. History
a. Know that a patient with an aspirated foreign body can be asymptomatic for hours or days after aspiration
2. Physical examination
a. Recognize the clinical presentation of foreign body aspiration
b. Recognize auscultatory findings typical of foreign body aspiration
3. Imaging
a. Describe the imaging techniques used in the diagnosis of foreign body aspiration (inspiratory-expiratory chest films, lateral decubitus chest films)
b. Recognize radiologic findings typical of aspirated foreign body
c. Recognize the limitations of imaging techniques in diagnosing foreign bodies
4. Pulmonary function tests
5. Other investigations
6. Other diagnostic criteria
a. Know the best methods for documenting chronic lipoid pneumonia in children
7. Complications
a. Recognize the chronic sequelae of retained aspirated foreign bodies

e. Prevention and therapeutic approach
    1. Prevention
    2. Therapeutic approach
        a. Know why chest physiotherapy is contraindicated in patients with foreign bodies in major airways
        b. Know the role of rigid endoscopy in the treatment of foreign body aspiration
        c. Know the therapy for hemoptysis associated with foreign body aspiration
        d. Recognize the indications for lobectomy to treat foreign body aspiration
        e. Know the immediate therapy for tracheal foreign body aspiration
        f. Know the treatment of lipoid pneumonia in children
    3. Side effects of therapy
f. Prognosis
    1. Natural history
    2. Prognosis with therapy

2. Aspiration of liquid and food
    a. Epidemiology
        1. Know the disease states most likely associated with aspiration of food or liquid in children
        2. Know the circumstances that predispose children to liquid aspiration
        3. Know that bottle propping is associated with the risk of lower airway disease
    b. Etiology/genetics
        1. Recognize the anatomic abnormalities that lead to recurrent pulmonary aspiration of liquids or foods
        2. Know that swallowing disorders in the absence of anatomic malformations can lead to recurrent aspiration of liquids and foods
        3. Know the association between brain stem abnormalities and swallowing dysfunction
    c. Pathophysiology
        1. Pathology
            a. Recognize the pathologic features characteristic of lipid aspiration in the lungs
            b. Recognize that chronic aspiration of lipid can lead to interstitial pulmonary fibrosis
        2. Path mechanisms and consequences
            a. Know that pH and volume of substance aspirated are major determinants of the severity of the lung injury in aspiration syndromes
            b. Understand the pathophysiologic consequences of massive aspiration of gastric contents
            c. Recognize the role of poor oral hygiene and gingivitis as risks in suppurative lung diseases
            d. Recognize the bacterial flora associated with pulmonary infections following aspiration of liquids and foods
    d. Diagnosis and clinical manifestations
        1. History
            a. Know that aspiration of ingested food or liquids can occur without acute coughing or choking

b. Recognize the clinical manifestations of chronic aspiration of liquids and foods in infants and children

2. Physical examination
   a. Recognize the physical findings associated with aspiration
3. Imaging
   a. Know that diagnostic findings may not appear on an x-ray study of the chest for several hours after an episode of aspiration
   b. Recognize the radiographic manifestations of acute and chronic aspiration of liquids and foods into the lungs
   c. Know the limitations of barium swallow in diagnosing recurrent aspiration of liquids and foods
   d. Understand the role of a multitexture video fluoroscopic swallowing study in the evaluation of swallowing dysfunction
   e. Recognize the relationship between swallowing dysfunction and the risk for chronic recurrent aspiration
4. Pulmonary function tests
5. Other investigations
6. Diagnostic criteria
7. Complications
   a. Recognize the pulmonary complications associated with chronic aspiration of liquids and foods

e. Prevention and therapeutic approach
   1. Prevention
   2. Therapeutic approach
      a. Recognize the indications for placement of a gastrostomy tube and fundoplication in the management of recurrent aspiration
      b. Know the therapies to employ in a child with primary swallowing disorder who experiences recurrent aspiration of liquid and food
      c. Know the therapy for lipid aspiration pneumonia
   3. Side effects of therapy
f. Prognosis
   1. Natural history
      a. Know the natural history of pulmonary disease in children with recurrent aspiration of liquids and foods
   2. Prognosis with therapy

3. Aspiration of hydrocarbons
   a. Epidemiology
      1. Identify common household products that put children at risk for hydrocarbon ingestion
      2. Know the risks of giving mineral oil to infants and neurologically impaired individuals
   b. Etiology/genetics
   c. Pathophysiology
      1. Pathology
      2. Path mechanisms and consequences

a. Understand the pathophysiology of pulmonary injury due to hydrocarbon ingestion

d. Diagnosis and clinical manifestations
   1. History
      a. Recognize the clinical features of hydrocarbon aspiration
   2. Physical examination
      a. Recognize findings typical of hydrocarbon aspiration on physical examination of the chest
   3. Imaging
      a. Know that pneumatoceles may be visible on x-ray study of the chest after hydrocarbon aspiration
      b. Understand the progression of radiographic changes following hydrocarbon aspiration
   4. Pulmonary function tests
   5. Other investigations
   6. Diagnostic criteria
   7. Complications
      a. Anticipate the immediate and long-term complications associated with hydrocarbon aspiration

e. Prevention and therapeutic approach
   1. Prevention
   2. Therapeutic approach
      a. Know that corticosteroids are not indicated in the treatment of hydrocarbon aspiration
      b. Know that the management of hydrocarbon pneumonitis in children is primarily supportive
   3. Side effects of therapy

f. Prognosis
   1. Natural history
      a. Know the progression of clinical features of hydrocarbon aspiration
      b. Know the natural history of acute hydrocarbon pneumonia
   2. Prognosis with therapy

4. Smoke inhalation
   a. Epidemiology
   b. Etiology/genetics
   c. Pathophysiology
      1. Pathology
         a. Know the pulmonary pathology of smoke inhalation injury
      2. Path mechanisms and consequences
         a. Understand the effects of inhalation of smoke from different types of fires (eg, flash electrical versus slower wood fires; burning plastics)
         b. Know the components of combustion that injure the lungs
         c. Recognize the immediate pathophysiologic changes associated with smoke inhalation

d. Know the effect of carbon monoxide poisoning (change in oxyhemoglobin dissociation curve) and the consequences of that change on unloading of oxygen in the peripheral tissues
e. Recognize the role of asphyxia in addition to that of lung injury in the outcome of victims of smoke inhalation

d. Diagnosis and clinical manifestations
1. History
a. Identify the clinical circumstances that predispose patients to carbon monoxide poisoning
2. Physical examination
a. Recognize the physical findings associated with carbon monoxide poisoning
b. Recognize the physical findings associated with an upper airway/laryngeal burn
c. Recognize the pulmonary physical findings associated with smoke inhalation
3. Imaging
a. Recognize the manifestations of smoke inhalation on x-ray study of the chest
4. Pulmonary function tests
a. Recognize the changes in gas exchange and lung mechanics associated with lung injury due to smoke inhalation
b. Know the blood concentrations of carbon monoxide that produce clinical problems and risk of death
5. Other investigations
6. Diagnostic criteria
7. Complications
a. Recognize the pulmonary and extrapulmonary complications associated with smoke inhalation

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
a. Know when to establish an airway in a patient with smoke inhalation and airway burns
b. Understand the rationale for 100% oxygen therapy for carbon monoxide poisoning
3. Side effects of therapy

f. Prognosis
1. Natural history
2. Prognosis with therapy

5. Near-drowning
a. Epidemiology
b. Etiology/genetics
c. Pathophysiology
1. Pathology
2. Path mechanisms and consequences

        a. Know that the amount of aspirated water is relatively small in most drowning victims and that approximately 15% die without aspiration
        b. Understand the pathogenesis of hypoxemia in near-drowning
        c. Understand the effect of hypothermia on the interpretation of blood gas values and acid-base status in near-drowning
        d. Understand the influence of hypothermia on the clinical manifestations associated with near-drowning
d. Diagnosis and clinical manifestations
    1. History
        a. Recognize historical factors that portend good outcomes despite near- drowning
    2. Physical examination
        a. Recognize the physical findings associated with near-drowning
    3. Imaging
        a. Know that hypoxemia may precede radiographic changes in near-drowning
    4. Pulmonary function tests
    5. Other investigations
        a. Know the electrolyte abnormalities associated with near-drowning
    6. Diagnostic criteria
    7. Complications
e. Prevention and therapeutic approach
    1. Prevention
    2. Therapeutic approach
        a. Understand the indications for and applications of supplemental oxygen and positive end-expiratory pressure in the management of near-drowning
        b. Know that the clinical condition on presentation influences the prognosis with therapy in near-drowning/drowning victims
    3. Side effects of therapy
f. Prognosis
    1. Natural history
    2. Prognosis with therapy
g. Gastroesophageal reflux
    1. Epidemiology
    2. Etiology
        a. Know the effects of increased respiratory work on gastroesophageal reflux
        b. Know that gastroesophageal reflux is more common in children with neuromuscular disorders and obstructive airway diseases than in healthy children
    3. Pathophysiology
        a. Know that gastroesophageal reflux may be associated with respiratory findings of cough and wheeze in the absence of pulmonary aspiration
        b. Know that esophageal irritation and distention can produce airway secretions via cholinergic neural reflexes
        c. Know the physiologic conditions that predispose to gastroesophageal reflux
        d. Know the pharmacologic agents that lower esophageal sphincter tone and predispose to gastroesophageal reflux

e. Know that transient increases in abdominal pressure can exceed normal lower esophageal sphincter tone, producing gastroesophageal reflux
f. Know that gastroesophageal reflux is associated with laryngospasm in some infants
g. Know that gastroesophageal reflux may be associated with laryngospasm

4. Diagnosis and clinical manifestations
   a. History
      1. Know that gastroesophageal reflux can produce respiratory symptoms in the absence of vomiting
   b. Physical findings
      1. Know the respiratory findings associated with gastroesophageal reflux in infants and children
   c. Imaging
      1. Know the limitations of imaging studies in gastroesophageal reflux
   d. Pulmonary function tests
   e. Other investigations
      1. Know the indications and interpretation of esophageal pH and impedance probe studies
      2. Know the limitations of esophageal pH and impedance probe studies in the diagnosis of pulmonary aspiration
   f. Other diagnostic criteria
      1. Know that the correlation between the esophageal pH and impedance probe findings and risk of lung disease in infants is poorly defined
   g. Complications
5. Prevention
   a. Prevention
   b. Therapeutic approach
      1. Know the therapies used to treat gastroesophageal reflux
   c. Side effects of therapy
      1. Know the complications of pharmacologic agents used to treat gastroesophageal reflux in children
6. Prognosis

L. Bronchopulmonary dysplasia
   1. Epidemiology
   2. Etiology/genetics
      a. Know the factors that are associated with an increased incidence of bronchopulmonary dysplasia
   3. Pathophysiology
      a. Pathology
         1. Recognize the typical gross and microscopic pathologic picture of lungs affected by bronchopulmonary dysplasia
         2. Know that older infants with bronchopulmonary dysplasia have enlarged alveolar air spaces on histologic examination
      b. Path mechanisms and consequences
         1. Know that compared to full-term infants, prematurely born infants have lower lung levels of anti-oxidant enzymes

2. Identify endothelial and epithelial cell functions that are impaired in patients with oxygen toxicity and know the time frame of this dysfunction
3. Know that prolonged exposure to high concentrations of oxygen or volutrauma can result in increased alveolar-capillary membrane permeability to solutes
4. Know that the higher compliance of the airways in a prematurely born infant may accentuate barotrauma and volutrauma injury
5. Know that bronchopulmonary dysplasia is associated with excess fluid administration or fluid retention in the newborn period
6. Know that hypertrophy of bronchial smooth muscle develops in infants with bronchopulmonary dysplasia
7. Know that hypertrophy of pulmonary artery smooth muscle develops in infants with bronchopulmonary dysplasia
8. Know that the appearance of leukocytes and their proteolytic enzymes in tracheobronchial fluids is associated with the evolution of idiopathic respiratory distress syndrome to bronchopulmonary dysplasia
9. Know the pathophysiologic consequences of airway obstruction in infants with bronchopulmonary dysplasia
10. Know that premature birth is associated with altered airway structure favoring obstruction, even in the absence of bronchopulmonary dysplasia

4. Diagnosis and clinical manifestations
   a. History
      1. Know that bronchopulmonary dysplasia can occur in infants of any gestational age who have neonatal lung injury
   b. Physical examination
      1. Know that recurrent wheezing frequently occurs in patients with bronchopulmonary dysplasia
   c. Imaging
      1. Recognize the features of bronchopulmonary dysplasia on imaging studies of the chest
      2. Know that during the first month after birth, chlamydial pneumonitis may mimic the picture of bronchopulmonary dysplasia on radiographic study of the chest
   d. Pulmonary function tests (see Prognosis)
      1. Recognize pulmonary function abnormalities in patients with bronchopulmonary dysplasia
   e. Other investigations
   f. Diagnostic criteria
      1. Know the diagnostic criteria for bronchopulmonary dysplasia
   g. Complications
      1. Know that infants with bronchopulmonary dysplasia frequently have a reversible component to their airway obstruction
      2. Know that children who had bronchopulmonary dysplasia as infants frequently have electrocardiograms compatible with right ventricular hypertrophy
      3. Know that tracheobronchomalacia is a complication of bronchopulmonary dysplasia
5. Prevention and therapeutic approach

a. Prevention
    1. Know the effects of surfactant replacement therapy on the incidence and severity of bronchopulmonary dysplasia
b. Therapeutic approach
    1. Know the effects of supplemental oxygen therapy on growth and pulmonary vascular pressures
    2. Know that infants with bronchopulmonary dysplasia frequently require higher-than-usual caloric intakes
    3. Understand the value and risks of administering corticosteroids systemically to facilitate extubation in ventilator-dependent infants with bronchopulmonary dysplasia
    4. Know that diuretics can be beneficial in patients with bronchopulmonary dysplasia
    5. Understand the influence of feeding or sleep state on systemic oxygenation in infants with bronchopulmonary dysplasia
    6. Differentiate ventilatory strategies that are appropriate in the treatment of bronchopulmonary dysplasia from those that are effective in patients with hyaline membrane disease
    7. Understand the concept of "controlled hypoventilation" in the management of patients with bronchopulmonary dysplasia who are receiving assisted ventilation
c. Side effects of therapy
    1. Recognize the side effects of chronic corticosteroid therapy in infancy
    2. Recognize the side effects of diuretic therapy in patients with bronchopulmonary dysplasia (i.e., metabolic alkalosis, hypokalemia, hypercalciuria, osteopenia)

6. Prognosis
a. Natural history
b. Prognosis with therapy
    1. Know that school-age children who had bronchopulmonary dysplasia as infants can have evidence of airflow limitation and air trapping on pulmonary function tests
    2. Know that school-age children who had bronchopulmonary dysplasia as infants have a high incidence of bronchial hyperreactivity to methacholine by inhalation

M. Pulmonary vascular diseases in childhood
1. Cor pulmonale
a. Epidemiology
b. Etiology
    1. Know the conditions most commonly associated with the development of cor pulmonale at various ages
c. Pathophysiology
    1. Pathology
        a. Recognize the pathophysiologic features of the heart that characterize cor pulmonale
    2. Path mechanisms and consequences

a. Know that sustained pulmonary hypertension produces cor pulmonale
b. Know that intermittent hypoxemia, such as that occurring during sleep, can produce pulmonary hypertension and subsequent cor pulmonale
c. Recognize the changes in heart function that occur in patients with cor pulmonale before right-sided heart failure occurs

d. Diagnosis and clinical manifestations
1. History
a. Recognize elements of the history suggestive of right-sided congestive heart failure
2. Physical examination
a. Recognize the typical physical findings in patients with right-sided heart failure
3. Imaging
a. Recognize the value of Doppler echocardiography in the diagnosis of pulmonary hypertension
4. Pulmonary function tests
5. Other investigations
6. Diagnostic criteria
a. Recognize the electrocardiographic features of right ventricular hypertrophy
b. Recognize the echocardiographic features associated with pulmonary hypertension
7. Complications
a. Know that right-sided heart failure is a complication of cor pulmonale

e. Prevention and therapeutic approach
1. Prevention
a. Understand the role of oxygen therapy in preventing cor pulmonale in a patient with chronic lung disease
2. Therapeutic approach
a. Know the supportive treatment for heart failure due to cor pulmonale
b. Understand the role of long-term oxygen therapy in the treatment of patients with cor pulmonale
3. Side effects of therapy

f. Prognosis
1. Natural history
2. Prognosis with therapy
a. Know that cor pulmonale can be reversed with successful therapy for underlying lung disease, e.g., upper airway obstruction

2. Pulmonary edema
a. Epidemiology
b. Etiology/genetics
1. Recognize causes of pulmonary edema
2. Know that pulmonary edema can result from rapid expansion of atelectatic lung tissue
3. Know that pulmonary edema can occur due to postcapillary pulmonary venous obstruction

4. Know that pulmonary edema can occur due to altered pulmonary endothelial cell permeability characteristics
5. Know that pulmonary edema can occur at high altitudes

c. Pathophysiology
   1. Pathology
      a. Know that pulmonary edema fluid may be blood tinged
      b. Know how to distinguish between cardiogenic and permeability pulmonary edema
      c. Identify the forces that promote fluid influx from the capillary bed into the interstitial spaces
   2. Path mechanisms and consequences
      a. Know that pulmonary edema can produce a restrictive and/or obstructive defect measured by standard pulmonary function tests
      b. Understand that pulmonary edema can cause V\Q mismatch
      c. Understand that pulmonary edema can cause small airway obstruction and wheezing
      d. Know that impaired lymphatic drainage may be associated with pulmonary edema

d. Diagnosis and clinical manifestations
   1. History
      a. Recognize the symptoms associated with pulmonary edema
   2. Physical examination
      a. Recognize the physical findings associated with pulmonary edema
   3. Imaging
      a. Recognize the radiographic features of pulmonary edema
      b. Differentiate the radiographic pattern of cardiogenic pulmonary edema from pulmonary edema due to altered capillary permeability
   4. Pulmonary function tests
   5. Other investigations
      a. Recognize the role of Swan-Ganz catheter measurements in the evaluation of pulmonary edema
   6. Diagnostic criteria
   7. Complications
      a. Know that low oncotic pressure reduces the pulmonary vascular pressure at which pulmonary edema develops
      b. Know that pulmonary edema increases pulmonary vascular resistance
      c. Know that pulmonary edema increases airway resistance

e. Prevention and therapeutic approach
   1. Prevention
   2. Therapeutic approach
      a. Know the supportive therapy for pulmonary edema
      b. Know that positive end-expiratory pressure redistributes but does not reduce lung water content in patients with pulmonary edema

f. Prognosis
   1. Natural history
   2. Prognosis with therapy

3. Pulmonary emboli/infarction
    a. Epidemiology
        1. Recognize the clinical factors that predispose a patient to pulmonary thromboembolism
    b. Etiology/genetics
        1. Recognize the clinical entities associated with pulmonary thrombosis
        2. Identify the causes of pulmonary emboli in children
        3. Recognize the genetic diseases that predispose children to pulmonary emboli
    c. Pathophysiology
        1. Pathology
            a. Recognize histologic features of pulmonary infarction
            b. Recognize the pathologic changes resulting from a large pulmonary embolus
        2. Path mechanisms and consequences
            a. Know that recurrent thromboembolic events can lead to pulmonary hypertension
            b. Understand the changes in gas exchange that occur immediately following pulmonary thromboembolism
            c. Understand the hemodynamic response to acute pulmonary embolism
            d. Understand that lung ischemia followed by reperfusion may produce free radicals and cause further lung injury
            e. Understand the role of bronchial blood flow during pulmonary embolism in maintaining perfusion to lung tissue distal to the embolus
    d. Diagnosis and clinical manifestations
        1. History
            a. Recognize the clinical features of pulmonary infarction
            b. Recognize the clinical features of massive pulmonary embolus involving large pulmonary vessels
        2. Physical examination
            a. Recognize the physical findings associated with massive pulmonary embolus
        3. Imaging
            a. Know the diagnostic techniques used to document the presence of pulmonary vascular obstruction due to emboli
            b. Recognize the value of spiral (helical) computed tomography with contrast in the diagnosis of a pulmonary embolus
        4. Pulmonary function tests
            a. Recognize the effect of pulmonary embolism on end-tidal PCO2 compared to arterial PCO2
        5. Other investigations
        6. Diagnostic criteria
        7. Complications
    e. Prevention and therapeutic approach
        1. Prevention
            a. Know the role of low molecular weight heparin in the prevention of pulmonary embolism in immobilized patients
        2. Therapeutic approach

3. Side effects of therapy

f. Prognosis
   1. Natural history
   2. Prognosis with therapy

4. Pulmonary hypertension
   a. Epidemiology
      1. Recognize the clinical conditions that predispose a child to pulmonary hypertension
   b. Etiology/genetics
      1. Know the role of bone morphogenic protein receptor 2(BMP-R2)
   c. Pathophysiology
      1. Pathology
         a. Recognize the features of structural remodeling of the pulmonary vascular bed associated with pulmonary hypertension due to chronic hypoxemia
         b. Know the pathophysiologic consequences of persistent pulmonary hypertension of the newborn
      2. Path mechanisms and consequences
         a. Recognize the gas exchange abnormalities associated with pulmonary hypertension
         b. Recognize the effects of pulmonary hypertension on cardiac function at rest
   d. Diagnosis and clinical manifestations
      1. History
         a. Recognize the symptoms associated with primary pulmonary hypertension and that most patients are asymptomatic
         b. Recognize the clinical features of conditions associated with pulmonary hypertension (chronic thromboembolic events, pulmonary fibrosis, congestive heart disease, etc.)
      2. Physical examination
         a. Recognize physical findings typical of primary pulmonary hypertension
      3. Imaging
         a. Recognize radiographic findings associated with primary pulmonary hypertension
      4. Pulmonary function tests
      5. Other investigations
      6. Diagnostic criteria
         a. Know the cardiac catheterization indices that separate pulmonary hypertension due to pulmonary vessel disease from that due to cardiac disorders
         b. Know the hemodynamic criteria for diagnosing pulmonary hypertension
      7. Complications
         a. Recognize the clinical complications associated with pulmonary hypertension
   e. Prevention and therapeutic approach
      1. Prevention
      2. Therapeutic approach

a. Understand the hemodynamic responses to pulmonary vasodilators in patients with pulmonary hypertension
b. Understand the role of oxygen in the treatment of pulmonary hypertension
c. Know the various pharmacologic agents used to treat pulmonary hypertension
d. Understand the mechanisms of action of different pharmacologic agents used to treat pulmonary hypertension

3. Side effects of therapy
a. Recognize dangers of pulmonary vasodilators in patients with pulmonary venous obstruction
b. Recognize the side effects of the pharmacologic agents used to treat pulmonary hypertension

f. Prognosis
1. Prognosis with therapy
a. Know that although the prognosis for primary pulmonary hypertension is poor, long-term survival has improved with newer pharmacologic treatments and that lung transplantation is a possible treatment

5. Pulmonary hemorrhage and hemosiderosis
a. Epidemiology
1. Recognize the causes of hemoptysis and pulmonary hemorrhage in childhood

b. Etiology/genetics
1. Know the differential diagnosis of diffuse alveolar hemorrhage in childhood
2. Understand that idiopathic pulmonary hemosiderosis is a diagnosis of exclusion
3. Understand that hemoptysis can result from focal or diffuse hemorrhage from airways or lung parenchyma
4. Understand that bleeding in the lung can arise from the high-pressure bronchial circulation or the low-pressure pulmonary circulation
5. Know the different diagnostic considerations between massive and minor hemoptysis

c. Pathophysiology
1. Pathology
a. Recognize the pulmonary histopathologic changes (including immunofluorescent findings) associated with Goodpasture syndrome
b. Recognize the pulmonary histopathologic features of pulmonary capillaritis
c. Recognize the pulmonary histopathologic features of idiopathic pulmonary hemosiderosis
d. Be able to recognize a hemosiderin-laden macrophage

2. Path mechanisms and consequences
a. Recognize the changes in bronchial circulation that occur due to bronchiectasis and understand how this may lead to hemoptysis
b. Understand the effects of pulmonary hemorrhage on gas exchange
c. Know the time course for the production of hemosiderin-laden macrophages

d. Diagnosis and clinical manifestations
1. History
a. Recognize clinical features suggestive of acute, recurrent, and chronic alveolar hemorrhage

b. Recognize the importance of travel history in the diagnosis of hemoptysis
c. Understand the importance of identifying other organ involvement in patients with diffuse alveolar hemorrhage syndromes
d. Recognize that retained foreign body is a common cause of massive hemoptysis in previously healthy children

2. Physical examination
   a. Recognize the clinical constellation of features characteristic of Goodpasture syndrome
   b. Recognize the physical findings associated with pulmonary hemorrhage
3. Imaging
   a. Recognize the radiographic features typical of diffuse alveolar hemorrhage
4. Pulmonary function tests
   a. Recognize the changes in pulmonary function associated with hemosiderosis (including diffusion capacity abnormalities)
5. Other investigations
   a. Understand the role of bronchoalveolar lavage in diagnosing alveolar hemorrhage
   b. Understand the limitations of identifying hemosiderin-laden macrophages in bronchoalveolar lavage in the diagnosis of alveolar hemorrhage syndromes
   c. Recognize the laboratory findings suggestive of immune- mediated lung disease in a child with pulmonary bleeding
   d. Recognize that a negative antineutrophil cytoplasmic antibody (ANCA) test does not exclude a diagnosis of pulmonary capillaritis
   e. Understand the role of cardiac and renal function testing and coagulation studies in evaluating patients with alveolar hemorrhage syndromes
   f. Understand the role of lung biopsy in the diagnosis of alveolar hemorrhage syndromes

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
   a. Know the treatment for massive pulmonary bleeding in a child
   b. Know the treatment for Goodpasture syndrome
   c. Know the current approaches to the treatment of a child with idiopathic pulmonary hemosiderosis
   d. Know the current approaches to the treatment for pulmonary capillaritis
3. Side effects of therapy
   a. Understand the risks associated with selective bronchial artery embolization in the treatment of pulmonary hemorrhage

f. Prognosis
1. Know the natural history of idiopathic pulmonary hemosiderosis is highly variable
2. Know the time course for radiographic resolution of pulmonary hemorrhage
3. Know the natural history of pulmonary bleeding associated with immune-mediated lung disease
4. Know the natural history of pulmonary hemorrhage associated with bronchiectasis in patients with cystic fibrosis

6. Pulmonary arteriovenous malformations
    a. Know the clinical presentation of pulmonary arteriovenous malformation and its association with hereditary hemorrhagic telangiectasia

N. Acute respiratory distress syndrome (ARDS)
1. Epidemiology
2. Etiology/genetics
    a. Recognize the clinical conditions associated with the development of acute respiratory distress syndrome in children
3. Pathophysiology
    a. Pathology
        1. Recognize the pathologic features of acute respiratory distress syndrome
    b. Path mechanisms and consequences
        1. Understand the pathophysiology of acute respiratory distress syndrome (including an increase in pro-inflammatory cytokines leading to increased alveolar capillary permeability)
        2. Know the mechanisms involved in the repair processes in patients with ARDS
        3. Know the conditions associated with an increased alveolar- arterial PO2 difference
        4. Know that the definition of ARDS includes normal left atrial pressure
        5. Know the role of epithelial sodium channels (ENaC) in alveolar fluid reabsorption
        6. Recognize the abnormalities in surfactant function and metabolism in patients with ARDS
4. Diagnosis and clinical manifestations
    a. History
        1. Know that acute respiratory distress syndrome can occur in neutropenic patients
    b. Physical examination
        1. Recognize the physical findings typical of acute respiratory distress syndrome
    c. Imaging
        1. Recognize the features of acute respiratory distress syndrome on x-ray study of the chest
    d. Pulmonary function tests
        1. Understand the changes in lung mechanics associated with acute respiratory distress syndrome
    e. Other investigations
        1. Know the effects of PEEP on pulmonary artery occlusion pressure measurements during hemodynamic monitoring in a patient with acute respiratory distress syndrome
    f. Diagnostic criteria
        1. Know that a ratio of PaO2/FIO2 < 300 defines acute lung injury and a PaO2/FIO2 < 200 reflects acute respiratory distress syndrome
    g. Complications
        1. Recognize complications associated with acute respiratory distress syndrome
5. Prevention and therapeutic approach
    a. Prevention
    b. Therapeutic approach

1. Know the supportive measures used to treat acute respiratory distress syndrome (eg, fluid therapy)
2. Know that prophylactic positive end-expiratory pressure does not prevent acute respiratory distress syndrome
3. Understand the effect of nitric oxide inhalation on the pulmonary vasculature
4. Know that PEEP can increase oxygenation in ARDS without decreasing the total lung water content
5. Recognize the lung-protective strategies (ventilation with low tidal volumes {6 mL/kg} and a plateau inspiratory pressure < 30 cm H2O) that improve outcomes in patients with established ARDS

c. Side effects of therapy
1. Understand that ventilation with high tidal volumes can increase circulating levels of inflammatory mediators that contribute to multiple organ system failure

6. Prognosis
a. Natural history
1. Know that multi-organ failure increases mortality associated with acute respiratory distress syndrome
2. Identify the modes of death associated with acute respiratory distress syndrome
3. Know the time course of pulmonary dysfunction in acute respiratory distress syndrome
4. Know that Gram-negative pneumonia and sepsis are frequent prodromal conditions in acute respiratory distress syndrome
5. Know the mortality associated with various causes of acute respiratory distress syndrome

b. Prognosis with therapy
1. Recognize the long-term pulmonary function abnormalities in survivors of acute respiratory distress syndrome

O. Pulmonary manifestations of immunosuppression
1. Acquired immunodeficiency syndrome
a. Epidemiology
1. Know the groups of patients that are at increased risk for the development of acquired immunodeficiency syndrome (AIDS) and AIDS-related lung disease

b. Etiology/Genetics
1. Know the common routes of human immunodeficiency virus infection in the neonatal and pediatric age groups

c. Pathophysiology
1. Pathology
a. Know that lymphocytic interstitial pneumonitis/pulmonary lymphoid hyperplasia are common forms of lung disease related to acquired immunodeficiency syndrome
b. Know that bronchiectasis can occur in patients with acquired immunodeficiency syndrome

2. Path mechanisms and consequences
a. Understand the alterations in lymphocyte subtype numbers that occur in patients with acquired immunodeficiency syndrome

d. Diagnosis and clinical manifestations
    1. History
    2. Physical examination
        a. Know that lymphocytic interstitial pneumonitis/pulmonary lymphoid hyperplasia can be associated with hepatosplenomegaly, clubbing, and parotid gland swelling
    3. Imaging
        a. Recognize lymphoid interstitial pneumonitis on radiographic study of the chest in the setting of HIV/AIDS
        b. Recognize Pneumocystis jiroveci (carinii) infection on radiographic study of the chest in the setting of HIV/AIDS
    4. Pulmonary function tests
    5. Other investigations
        a. Know the importance of excluding cardiomyopathy and pulmonary edema in a child with HIV/AIDS who has acute respiratory distress
        b. Know that IgG antibody-based diagnostic tests for HIV may reflect maternal antibody and not infection of the infant in the first months of life
        c. Know the role of bronchoalveolar lavage and induced sputum in the diagnosis of opportunistic infection
        d. Know the role of evaluating CD4 counts in assessing risk for infection and need for prophylaxis of Pneumocystis jiroveci (carinii)
    6. Diagnostic criteria
        a. Know the diagnostic criteria for acquired immunodeficiency syndrome (AIDS) and AIDS-related complex, as established by the Center for Disease Control
    7. Complications
        a. Know that atypical mycobacteria can cause pulmonary and disseminated disease in patients with immunodeficiency
        b. Know that HIV/AIDS patients are at increased risk for tuberculosis
        c. Know that HIV/AIDS patients are at risk for severe infections from common viral agents: herpes simplex, CMV, RSV, and other respiratory pathogens
        d. Know that neonatal HIV/AIDS patients are at risk for severe infections from common encapsulated bacteria: Streptococcus pneumoniae, Staphylococcus aureus, Haemophilus influenzae
        e. Know that HIV/AIDS patients are at risk for severe infections from fungi: Pneumocystis jiroveci (carinii), Candida albicans, Histoplasma capsulatum, Cryptococcus neoformans, Aspergillus species, Coccidioides immitis
e. Prevention and therapeutic approach
    1. Prevention
        a. Know the role of maternal anti-retroviral therapy and neonatal anti-retroviral therapy in reducing the risk of neonatal transmission of HIV
    2. Therapeutic approach
        a. Know the treatment of tuberculosis in patients with acquired immunodeficiency syndrome
        b. Recognize the indications for prophylaxis against Pneumocystis jiroveci (carinii) in patients with acquired immunodeficiency syndrome

c. Know the therapeutic agents and approaches that are used in prophylaxis against Pneumocystis jiroveci (carinii) in patients with acquired immunodeficiency syndrome
d. Know the antimicrobial agents that are useful in the treatment of acute Pneumocystis jiroveci (carinii) pneumonia in patients with acquired immunodeficiency syndrome
e. Know that corticosteroids are adjunctive therapeutic agents in patients with acquired immunodeficiency syndrome who have acute Pneumocystis jiroveci (carinii) pneumonia

3. Side effects of therapy
   a. Recognize the nonpulmonary side effects of antimicrobial agents used to treat patients with acquired immunodeficiency syndrome who have Pneumocystis jiroveci (carinii) pneumonia
   b. Know that bronchospasm caused by inhalation of pentamidine can be prevented by prior inhalation of albuterol

f. Prognosis
   1. Natural history
   2. Prognosis with therapy
      a. Know that disseminated atypical mycobacterial pulmonary infection in patients with acquired immunodeficiency syndrome is usually lethal despite antibacterial therapy
      b. Know that the prognosis of HIV/AIDS is significantly improved with use of highly active anti-retroviral treatment (HAART)

2. Miscellaneous immunosuppressive disorders
   a. Epidemiology
      1. Know that common variable immunodeficiency (CVID) is the most prevalent of the primary immunodeficiency diseases
   b. Etiology/genetics
      1. Know the main diagnostic categories of patients with immunosuppression at risk for lung infection: primary immunodeficiencies, immuno-suppressive therapy (systemic corticosteroids in autoimmune disease, cancer patients, transplant patients), and HIV/AIDS
   c. Pathophysiology
      1. Pathology
         a. Recognize the typical pathologic findings in patients with chronic granulomatous disease
         b. Know that T-cell immunodeficiency is associated with increased risk for intracellular pathogens (e.g., viruses, fungi, and mycobacteria)
         c. Know that T-cell immunodeficiency is associated with increased risk for severe infection from common respiratory viruses such as RSV and parainfluenza
         d. Know that patients with neutrophil, immunoglobulin, or complement deficiency are at increased risk for severe bacterial infections
      2. Path mechanisms and consequences

a. Know that neutrophils, monocytes, and alveolar macrophages in patients with chronic granulomatous disease have defective oxidative bursts and cannot produce oxygen radicals
b. Know which microorganisms cause morbidity in patients with chronic granulomatous disease
c. Know the immune defect(s) in severe combined immune deficiency
d. Know the microorganisms that cause morbidity in children with severe combined immune deficiency
e. Identify organisms that cause morbidity in patients with IgA deficiency
f. Know that 20% to 30% of patients with IgA deficiency have associated IgG subclass deficiency
g. Know that recurrent bacterial infections of upper and lower respiratory tracts are the main clinical manifestations of common variable immunodeficiency and that bronchiectasis may develop if optimal therapy is delayed
h. Know the common infective agents seen in common variable immunodeficiency: Haemophilus influenzae, Moraxella catarrhalis, Streptococcus pneumoniae, and Staphylococcus aureus

d. Diagnosis and clinical manifestations
1. History
a. Know that IgG deficiency is associated with chronic (recurrent) pulmonary infections
2. Physical examination
3. Imaging
4. Pulmonary function tests
a. Know that lung disease secondary to IgG or IgA deficiency usually has an obstructive ventilatory pattern on pulmonary function testing
5. Other investigations
a. Know how to establish the diagnosis of chronic granulomatous disease on the basis of laboratory data
b. Know that decreased concentrations of IgG2 and IgG4 are associated with recurrent pulmonary infections
c. Know that healthy patients can have decreased concentrations of IgG2 and IgG4
6. Diagnostic criteria
7. Complications
a. Know that the incidence of pneumococcal, Haemophilus influenzae and Salmonella pneumonias is increased in patients with sickle cell disease

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
3. Side effects of therapy
a. Know that patients with IgA deficiency may have anaphylactic reactions to intravenous immune globulin therapy
b. Know which vaccines can safely be administered to patients with immune deficiency

f. Prognosis
    1. Natural history
    2. Prognosis with therapy
        a. Know that chronic bronchitis and decreased airflow rates frequently develop in patients with agammaglobulinemia despite immunoglobulin replacement therapy

P. Control of breathing disorders
1. Obstructive sleep apnea syndrome
    a. Epidemiology
        1. Know the incidence of habitual snoring in children
        2. Know the relationship between snoring and obstructive sleep apnea syndrome
        3. Know that in children, the prevalence of obstructive sleep apnea syndrome is equal in prepubertal males and females
    b. Etiology/genetics
        1. Know that hypertrophy of tonsils and adenoids is the most common factor predisposing children to obstructive sleep apnea syndrome
        2. Know that the incidence of obstructive sleep apnea syndrome is increased in craniofacial syndromes associated with micrognathia and mid-face hypoplasia
        3. Know that morbid obesity is associated with obstructive sleep apnea syndrome, especially in adolescence
    c. Pathophysiology
        1. Pathology
            a. Know that closure of the upper airway is greater with a small upper airway; decreased upper airway skeletal muscle tone; and increased inspiratory force
        2. Path mechanisms and consequences
            a. Know that the hypopharynx is a frequent site of obstruction in obstructive sleep apnea syndrome
            b. Know that nasal congestion can increase upper airway resistance
            c. Know that children with marked hypoxemia and hypercapnia may have normal gas exchange while awake
            d. Know the impact of sleep on respiratory function and gas exchange in normal children with lung disease
            e. Know that both adenoidal and tonsillar hypertrophy can contribute to obstructive sleep apnea syndrome
    d. Diagnosis and clinical manifestations
        1. History
            a. Understand the difference between primary snoring and obstructive sleep apnea syndrome
            b. Recognize the clinical signs and symptoms of obstructive sleep apnea syndrome in children during sleep as well as during wakefulness
            c. Know the significance of a history of snoring
            d. Know that there is no correlation between the intensity (loudness) of snoring and the severity of obstructive sleep apnea syndrome in children
            e. Know that snoring in children is often continuous without evidence of apnea
            f. Know the importance of a history of poor school performance or behavioral problems in children with snoring

2. Physical examination
    a. Know that midface hypoplasia and craniofacial anomalies increase the likelihood of obstructive sleep apnea syndrome
3. Imaging
    a. Know that lateral neck radiographs (high KV or soft tissue exposure) are useful in assessing the size of adenoids, sublingual tonsils, and the upper airway
4. Pulmonary function tests
    a. Know that pulmonary function tests are often normal in children with obstructive sleep apnea syndrome
5. Other investigations
    a. Understand the limitations of audio and video taping a sleeping child as a screening test for obstructive sleep apnea syndrome
    b. Know what physiologic measurements are made on polysomnography in children
    c. Understand the limitations and advantages of monitoring respiration (effort and airflow) during sleep
    d. Understand the limitations of daytime nap studies in diagnosing obstructive sleep apnea syndrome in children
    e. Recognize the features of central and obstructive apnea on a polysomnogram
    f. Recognize the features of hypopnea on a polysomnogram
    g. Understand the limitations of monitoring oxyhemoglobin saturation as a means of detecting obstructive apnea in children
6. Diagnostic criteria
    a. Know the diagnostic criteria for establishing the presence of obstructive sleep apnea
7. Complications
    a. Know the complications of obstructive sleep apnea
    b. Recognize the association between sleep disorder breathing and school problems

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
    a. Know that sedation may worsen obstructive sleep apnea syndrome in children
    b. Know that adenotonsillectomy is the most common treatment of obstructive sleep apnea syndrome in children
    c. Know that a history of snoring is not by itself an indication for treatment
    d. Know that weight loss is an important part of the treatment of the obese adolescent with obstructive sleep apnea syndrome
    e. Recognize the indications for mask continuous positive airway pressure therapy in children and adolescents with obstructive sleep apnea syndrome
    f. Know that positive airway pressure can improve obstructive sleep apnea
3. Side effects of therapy

a. Know that the risk of postoperative respiratory complications is increased in children younger than 2 years of age who have severe obstructive sleep apnea syndrome, craniofacial abnormalities, and a history of premature birth
b. Recognize the complications of mask continuous positive airway pressure in the treatment of obstructive sleep apnea syndrome
c. Know that an adenotonsillectomy does not always relieve symptoms of obstructive sleep apnea syndrome, especially in those who are obese or who have craniofacial abnormalities

f. Prognosis
1. Natural history
2. Prognosis with therapy
a. Know that most children without underlying craniofacial disorders or obesity will have complete resolution of symptoms of obstructive sleep apnea syndrome following tonsillectomy and adenoidectomy

2. Congenital central hypoventilation syndrome
a. Epidemiology
1. Know that congenital central hypoventilation syndrome is a rare cause of respiratory failure in children
b. Genetics
1. Know that alveolar hypoventilation syndromes may be congenital, acquired, and/or transient
2. Know that certain metabolic and structural abnormalities affecting the central nervous system may contribute to central hypoventilation syndrome
3. Know that mutations of the PHOX2B gene cause congenital central hypoventilation syndrome
c. Pathophysiology
1. Pathology
2. Path mechanisms and consequences
a. Know that oxygen and carbon dioxide responses are abnormal in patients with congenital central hypoventilation syndrome
b. Know that congenital central hypoventilation syndrome is a generalized disorder of the autonomic nervous system
c. Know that congenital central hypoventilation syndrome can be associated with Hirschsprung disease and tumors of neural crest origin
d. Diagnosis and clinical manifestations
1. History
a. Recognize the diagnostic features of congenital central hypoventilation syndrome
b. Know the conditions (lung disease, heart disease, brain stem malformations, neuromuscular disease, and metabolic disorders) that should be excluded before making a diagnosis of congenital central hypoventilation syndrome
c. Know that some congenital central hypoventilation syndrome patients can present late as children or adults, usually with evidence of an abnormal ventilatory response under stress, such as failure to breathe after general anesthesia
2. Physical examination

a. Know that children with congenital central hypoventilation syndrome may have findings of dysrhythmias or alterations in blood pressure
b. Know that children with congenital central hypoventilation syndrome may demonstrate no signs of dyspnea or stress despite inadequate ventilation or pneumonia

3. Imaging
   a. Know that magnetic resonance imaging (MRI) is the investigation of choice to detect brainstem abnormalities
4. Other studies
   a. Recognize the indications for ventilatory response testing
   b. Understand the role of polysomnography in establishing a diagnosis of congenital central hypoventilation syndrome
5. Diagnostic criteria
   a. Know the diagnostic criteria (clinical and laboratory data) for establishing a diagnosis of an alveolar hypoventilation syndrome
   b. Know that increased intracranial pressure and/or brain stem compression can cause respiratory control abnormalities
   c. Know that mutations of the PHOX2B gene are diagnostic of congenital central hypoventilation syndrome
6. Complications
   a. Know that death due to hypoxemia during sleep is a complication of alveolar hypoventilation syndrome

e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
   a. Know the indications for and the effectiveness of the various therapeutic approaches to treatment of congenital central hypoventilation syndrome
   b. Recognize the impact of long-term ventilatory support on the child and family
   c. Identify the resources and skills necessary for successful home management of a child with congenital central hypoventilation syndrome
   d. Know that pharmacologic therapy is ineffective in the management of congenital central hypoventilation syndrome
   e. Recognize the indications for and the risks and benefits of diaphragmatic or phrenic nerve pacing in patients with congenital central hypoventilation syndrome
3. Side effects of therapy
   a. Recognize the complications of long-term ventilation (positive and negative pressure)
   b. Recognize the complications of diaphragmatic pacing

f. Prognosis
1. Natural history
   a. Know that the condition of children with congenital central hypoventilation syndrome most likely will not improve with age
2. Prognosis with therapy

a. Know that even with appropriate therapy, the long-term prognosis for children with congenital central hypoventilation syndrome is variable but the need for ventilatory support is life long
b. Know that the development of cor pulmonale in congenital central hypoventilation syndrome is presumed to be due to inadequate ventilation during sleep and/or wakefulness until proven otherwise

3. Apnea of prematurity
   a. Epidemiology
      1. Understand the relationship between gestational age and apnea of prematurity
      2. Understand that stresses can exacerbate apnea of prematurity, such as anemia, hypoglycemia, electrolyte imbalance, and sepsis
   b. Etiology/genetics
   c. Pathophysiology
      1. Pathology
         a. Know that apnea of prematurity may be central, obstructive, or mixed
      2. Path mechanisms and consequences
         a. Know that premature infants have a biphasic ventilatory response to hypoxia, with an initial increase in minute ventilation followed by a reduction in minute ventilation to baseline or below
   d. Diagnosis and clinical manifestations
      1. History
         a. Know that isolated bradycardia in an infant who was born prematurely is suggestive of hypoxemia
         b. Recognize underlying conditions associated with increased apnea in infants who were born prematurely
      2. Physical examination
         a. Recognize the clinical manifestations of apnea of prematurity
      3. Imaging
      4. Other studies
         a. Recognize the indications for polysomnography in infants with apnea of prematurity
         b. Understand impedance monitoring can detect central apnea but not hypoxemia or airway destruction in patients with apnea of prematurity
      5. Diagnostic criteria
      6. Complications
   e. Prevention and therapeutic approach
      1. Prevention
      2. Therapeutic approach
         a. Recognize the indications for therapy in patients with apnea of prematurity
         b. Recognize the indications for home monitoring in infants with apnea of prematurity
         c. Know that caffeine is used to treat apnea of prematurity
         d. Know that nasal CPAP is a nonpharmacologic method for treatment of apnea of prematurity
      3. Side effects of therapy
         a. Recognize the side effects of caffeine in infants with apnea of prematurity

f. Prognosis
    1. Natural history
        a. Know that the typical history of apnea of prematurity is complete resolution after the infant reaches term
    2. Prognosis with therapy

4. Apparent life-threatening events
    a. Epidemiology
        1. Know that an associated diagnosis cannot be found for an apparent life-threatening event in approximately 50% of the incidents
    b. Etiology/genetics
        1. Identify conditions associated with apparent life-threatening events
        2. Know that a minority of sudden infant death syndrome cases have a history of an apparent life-threatening event
    c. Pathophysiology
        1. Pathology
        2. Path mechanisms and consequences
    d. Diagnosis and clinical manifestations
        1. History
            a. In evaluating an apparent life-threatening event, recognize the importance of obtaining a complete history of the event from the caregiver who witnessed the incident
            b. Recognize the importance of the history in determining the need for therapy for an apparent life-threatening event
        2. Physical examination
            a. Recognize normal and abnormal breathing patterns in young infants
        3. Imaging
        4. Pulmonary function tests
        5. Other investigations
            a. Know that pneumograms and polysomnograms do not predict the risk of subsequent apparent life-threatening events
            b. Recognize the utility of obtaining a measure of acid-base status (pH, HCO3) quickly following an apparent life-threatening event
        6. Diagnostic criteria
            a. Know that the current definition of an apparent life-threatening event is not limited to apnea only
        7. Complications
    e. Prevention and therapeutic approach
        1. Prevention
        2. Therapeutic approach
            a. Know the importance of extensive parental education in monitoring techniques and cardiopulmonary resuscitation for infants who have had apparent life-threatening events and are being monitored
        3. Complications of therapy
            a. Understand the limitations of home monitoring for apparent life-threatening events
            b. Recognize the consequences of home monitoring on care givers

f. Prognosis
    1. Natural history
    2. Prognosis with therapy
        a. In an infant who has experienced an apparent life-threatening event, know that home monitoring may not prevent death in a subsequent episode

5. Sudden infant death syndrome
    a. Epidemiology
        1. Know the current definition of sudden infant death syndrome
        2. Know the incidence of sudden infant death syndrome in the general population
        3. Know the characteristic age-related distribution of sudden infant death syndrome
        4. Recognize the risk factors for sudden infant death syndrome
        5. Understand the relationship between supine position and sudden infant death syndrome
    b. Etiology/genetics
        1. Know that the risk of sudden infant death syndrome (SIDS) is not significantly increased in first-degree relatives of a SIDS victim
    c. Pathophysiology
        1. Pathology
            a. Recognize the pathologic markers thought to be characteristic of sudden infant death syndrome
        2. Path mechanisms and consequences
    d. Diagnosis and clinical manifestations
        1. History
            a. Understand the importance of obtaining a complete history from the caregivers of an infant who died of sudden infant death syndrome
            b. Understand the importance of a "death scene" investigation in evaluating a case of sudden infant death syndrome
            c. Understand the importance of obtaining a family history in evaluating a case of sudden infant death syndrome
            d. Know that the possibility of child abuse should be considered in a child if more than one sibling has previously died, apparently of sudden infant death syndrome
        2. Physical examination
        3. Imaging
        4. Pulmonary function tests
        5. Other investigations
        6. Diagnostic criteria
            a. Know that the determination of sudden infant death syndrome must include autopsy
        7. Complications
    e. Prevention and therapeutic approach
        1. Prevention
            a. Understand that maternal smoking both during and after pregnancy increases the risk of sudden infant death syndrome

b. Know that the incidence of SIDS has decreased after public health campaigns to: have infants sleep on their backs; remove soft bedding, pillows, quilts, and stuffed animals from cribs; decrease environmental tobacco smoke; avoid overheating of infants

2. Therapeutic approach
   a. Know the appropriate management of the grieving family of an infant who died of sudden infant death syndrome
3. Side effects of therapy

f. Prognosis
1. Natural history
2. Prognosis with therapy

Q. Miscellaneous lung diseases
1. Alpha1-Protease Inhibitor Deficiency
   a. Epidemiology
      1. Recognize the association between cigarette smoking and symptomatic alpha1-protease inhibitor deficiency
   b. Etiology/genetics
      1. Know that alpha1-protease inhibitor deficiency is inherited in an autosomal recessive fashion
   c. Pathophysiology
      1. Pathology
         a. Know that alpha1-protease inhibitor deficiency is associated with the development of emphysema, asthma, and chronic airway inflammation
      2. Path mechanisms and consequences
         a. Understand the protease/antiprotease theory of lung disease associated with alpha1-protease inhibitor deficiency
         b. Understand the pathophysiology of liver disease in patients with alpha1-protease inhibitor deficiency
   d. Diagnosis and clinical manifestations
      1. History
         a. Know that the typical age of onset of pulmonary symptoms caused by alpha1- protease inhibitor deficiency is the third or fourth decade
      2. Physical examination
      3. Imaging
      4. Pulmonary function tests
         a. Recognize the pulmonary function test findings typical of emphysema
      5. Other investigations
         a. Know the significance of the following phenotypes: ZZ, SS, MM, MZ, and NULL
      6. Diagnostic criteria
      7. Complications
   e. Prevention and therapeutic approach
      1. Prevention
         a. Know that the onset of symptoms due to alpha1-protease inhibitor deficiency is greatly accelerated in lifelong smokers

b. Know that smoking causes oxidant inactivation of intrapulmonary proteinase inhibitors
2. Therapeutic approach
a. Recognize the indications for replacement therapy in patients with alpha1-protease inhibitor deficiency
3. Side effects of therapy
f. Prognosis
1. Natural history
2. Prognosis with therapy

2. Ciliary dysmotility
a. Epidemiology
b. Etiology/genetics
1. Know the mode of inheritance of ciliary dysmotility
c. Pathophysiology
1. Pathology
a. Know the ultrastructural defects of cilia in patients with ciliary dysmotility
2. Path mechanisms and consequences
a. Know how ciliary dysmotility leads to airways disease
b. Understand the relationship between ciliary dysmotility and male sterility
d. Diagnosis and clinical manifestations
1. History
a. Recognize the features of ciliary dysmotility in children
b. Know that neonatal respiratory disorders occur in the majority of children with ciliary dysmotility
2. Physical examination
3. Imaging
a. Understand the association between situs inversus and ciliary dysmotility
4. Pulmonary function tests
5. Other investigations
a. Recognize the indications for ciliary biopsy and/or sperm analysis
b. Know approaches to the measurement of ciliary transport rates and ciliary beat frequency
c. Know the value of nasal nitric oxide in the diagnosis of ciliary dysmotility
6. Diagnostic criteria
a. Know the diagnostic criteria for ciliary dysmotility
b. Recognize the limitations of ciliary biopsy in establishing the diagnosis of ciliary dysmotility following an acute viral infection
7. Complications
e. Prevention and therapeutic approach
1. Prevention
2. Therapeutic approach
f. Prognosis
1. Natural history
a. Know the natural history of ciliary dysmotility
2. Prognosis with therapy

3. Oncology

a. Epidemiology
b. Etiology/genetics
c. Pathophysiology
    1. Pathology
        a. Know the primary tumors of the respiratory system during childhood
        b. Identify the tumors that metastasize to the lung in children
        c. Know which lymph nodes drain the right and left lungs
    2. Path mechanisms and consequences
d. Diagnosis and clinical manifestations
    1. History
    2. Physical examination
    3. Imaging
        a. Be able to localize tumors on computed tomography (CT scan) of the chest
    4. Pulmonary function tests
    5. Other investigations
    6. Diagnostic criteria
    7. Complications
e. Prevention and therapeutic approach
    1. Prevention
    2. Therapeutic approach
    3. Side effects of therapy
        a. Recognize the adverse pulmonary side effects of chemotherapeutic drugs
f. Prognosis
    1. Natural history
    2. Prognosis with therapy

4. Trauma
    a. Know that the majority of chest trauma injuries in children are caused by blunt trauma sustained during an automobile accident
    b. Know that in children (as opposed to adults), serious intrathoracic injury often occurs in the absence of obvious chest wall injury (eg, rib fractures) following blunt trauma
    c. Know the common consequences of severe blunt chest trauma (eg, pneumothorax, hemothorax, airway obstruction, cardiac tamponade, flail chest, tracheobronchial tears, pulmonary contusion, ruptured diaphragm)
    d. Recognize the physical findings characteristic of a flail chest
    e. Know the appropriate management of a flail chest
    f. Recognize the clinical features of pulmonary contusion
    g. Know the natural history of pulmonary contusion
    h. Recognize the radiologic manifestations of pulmonary contusion
    i. Recognize the signs and symptoms of a traumatic tracheobronchial rupture
    j. Know the diagnostic and therapeutic options for a traumatic tracheobronchial tear (eg, jet or oscillatory ventilation)
    k. Know that following fractures of long bones, fat embolism may cause adult respiratory distress syndrome
    l. Know that the classic clinical triad of fat embolism includes respiratory insufficiency, neurologic dysfunction, and petechiae

m. Know the clinical manifestations of traumatic spinal cord injury
n. Recognize the signs and symptoms of pulmonary embolism following long bone fracture

5. Sarcoidosis
   a. Epidemiology
   b. Etiology/genetics
   c. Pathophysiology
      1. Pathology
         a. Be able to recognize a non-caseating granuloma
      2. Path mechanisms and consequences
         a. Know the pathogenesis of sarcoidosis
   d. Diagnosis and clinical manifestations
      1. History
         a. Recognize the typical pulmonary and nonpulmonary clinical manifestations of sarcoidosis
      2. Physical examination
         a. Know the importance of slit-lamp examination in patients with sarcoidosis
      3. Imaging
         a. Recognize the findings typical of sarcoidosis on x-ray study of the chest
      4. Pulmonary function tests
         a. Recognize the pulmonary function test findings typical of sarcoidosis
      5. Other investigations
         a. Know that elevation of serum angiotensin-converting enzyme activity is not specific for sarcoidosis
         b. Recognize findings typical of sarcoidosis on bronchoalveolar lavage
         c. Know the laboratory studies that are abnormal in sarcoidosis (eg, serum calcium, ESR, immunoglobulins, angiotensin-converting enzyme, leukopenia, or eosinophilia)
      6. Diagnostic criteria
         a. Recognize that the diagnosis of sarcoidosis is best established by biopsy of an affected lymph node
   e. Prevention and therapeutic approach
      1. Prevention
      2. Therapeutic approach
         a. Recognize the absolute indications for corticosteroid treatment in patients with sarcoidosis
      3. Side effects of therapy
   f. Prognosis
      1. Natural history
         a. Know the usual natural history of sarcoidosis in children
      2. Prognosis with therapy

6. Hypersensitivity pneumonitis
   a. Epidemiology
   b. Etiology/genetics
      1. Know that hypersensitivity pneumonitis is caused by inhalation of organic dusts
      2. Know the most common antigens associated with hypersensitivity pneumonitis

c. Pathophysiology
    1. Pathology
    2. Path mechanisms and consequences
        a. Know that the pathogenesis of hypersensitivity pneumonitis involves IgG-mediated and lymphocyte-mediated mechanisms but not IgE-mediated mechanisms
d. Diagnosis and clinical manifestations
    1. History
        a. Know that hypersensitivity pneumonitis may manifest itself as an acute, subacute, or chronic illness
        b. Recognize the historical features typical of hypersensitivity pneumonitis
    2. Physical examination
        a. Know that inspiratory crackles, not wheezes, are characteristic of acute hypersensitivity pneumonitis
    3. Imaging
        a. Recognize the radiographic manifestations of hypersensitivity pneumonitis
    4. Pulmonary function tests
    5. Other investigations
        a. Know that patients with hypersensitivity pneumonitis will have precipitating antibodies to the offending antigen but that the presence of such antibodies alone is not diagnostic
        b. Recognize findings typical of hypersensitivity pneumonitis on bronchoalveolar lavage
    6. Diagnostic criteria
    7. Complications
        a. Know that hypersensitivity pneumonitis may progress to end-stage pulmonary fibrosis and respiratory failure
e. Prevention and therapeutic approach
    1. Prevention
    2. Therapeutic approach
    3. Side effects of therapy
f. Prognosis
    1. Natural history
        a. Know that the condition of most patients with hypersensitivity pneumonitis will improve after they are removed from the source of antigen
    2. Prognosis with therapy

7. Wegener granulomatosis
    a. Know which therapy is most likely to induce remission of Wegener granulomatosis
8. Neuromuscular disorders
    a. Understand that patients with Duchenne muscular dystrophy usually have normal ventilatory drive
    b. Understand how chest wall compliance is affected by chronic neuromuscular disorders
    c. Recognize the inheritance pattern and genetic mechanism underlying Duchenne muscular dystrophy (X-linked recessive loss of function)

d. Recognize the inheritance pattern and genetic mechanism underlying spinal muscular atrophy
e. Recognize that sleep-disordered breathing typically precedes overt respiratory failure in patients with neuromuscular disease
f. Recognize that impaired airway clearance is a distinct risk factor for morbidity and mortality in neuromuscular disorders

9. Pulmonary alveolar proteinosis
   a. Recognize that pulmonary alveolar proteinosis can be congenital or acquired
   b. Understand the role of GM-CSF in surfactant homeostasis
   c. Know that the usual therapy for symptomatic pulmonary alveolar proteinosis is whole lung lavage
10. Prader-Willi syndrome
   a. Know that children with Prader-Willi syndrome also have an abnormal ventilatory response to hypoxemia
   b. Know that obese children with Prader-Willi syndrome have a decreased ventilatory response to hypercapnia and hypoxia
11. Familial dysautonomia
   a. Know that patients with familial dysautonomia (Riley-Day syndrome) have a reduced or absent ventilatory response to hypoxemia

2. **Evaluation/Diagnosis**

A. Pulmonary history
   1. Understand the importance of environmental exposure to animals, allergens, cigarette smoke, wood-burning stoves, and air pollutants in various respiratory diseases
   2. Recognize the effect of immunizations on the risk and severity of subsequent respiratory infections
   3. Identify the symptoms that assist in diagnostic evaluation of chronic chest pain (chronic cough, acute/chronic wheeze, hemoptysis, dyspnea) in childhood
   4. Know that globus hystericus, hyperventilation syndrome, chronic nonproductive cough, chest pain, and laryngeal wheezing may have psychological components
   5. Know the differential diagnosis of acute and chronic cough
   6. Know that psychogenic or "habit" cough is characterized by its stereotypical nature and improvement during sleep

B. Physical examination
   1. Inspection
      a. Cyanosis
         1. Know that peripheral cyanosis is a common finding in healthy children without hypoxemia
         2. Understand the relationship between cyanosis and the concentration of desaturated hemoglobin
         3. Understand the influence of carboxyhemoglobin and methemoglobin on skin color
      b. Respiratory pattern
         1. Know the normal range of respiratory rate by age
         2. Recognize abnormal respiratory patterns, including periodic breathing and Cheyne-Stokes, Biot, and Kussmaul respiration
         3. Recognize periodic breathing

4. Recognize the effects of inspiratory and expiratory obstruction on inspiratory to expiratory ratio

c. Retractions/bulging
1. Know that intercostal and suprasternal retractions, nasal flaring, and increased rate and depth of respiration indicate increased respiratory drive and increased work of breathing
2. Know that chest wall retractions usually reflect abnormally negative intrapleural pressure and/or a highly compliant chest wall

d. Chest size and shape
1. Recognize abnormal chest wall configurations, including pectus excavatum, pectus carinatum, barrel chest, scoliosis, kyphosis, “bell-shape" asphyxiating thoracic dystrophy, and rickets
2. Know the definition and differential diagnosis of an increase in the anteroposterior diameter of the chest

e. Diaphragmatic function
1. Recognize the appearance of asymmetric diaphragmatic activity

f. Digital clubbing/pulmonary osteoarthropathy
1. Identify diseases that predispose patients to digital clubbing/pulmonary osteoarthropathy
2. Recognize the characteristics of early digital clubbing

2. Percussion
a. Understand the clinical significance of changes in percussion (dullness vs tympany vs hyper-resonance)

3. Auscultation
a. Know the projections of the bronchopulmonary segments on the chest wall
b. Recognize and differentiate stridor, crackles, wheezes, pleural and pericardial friction rubs
c. Understand the pathophysiologic mechanisms responsible for crackles and wheezes

4. Palpation
a. Recognize the causes of a deviated cervical trachea
b. Recognize the importance of costochondral tenderness
c. Recognize the significance of crepitus over the chest wall

5. General
a. Know that pulsus paradoxus reflects the change in pleural pressure between inspiration and expiration
b. Recognize the importance of a loud pulmonic valve closure sound
c. Understand the importance of examining the ear canals in a patient with chronic cough

C. Pulmonary function testing
1. Static lung volumes
a. Spirometry
1. Know that lung volumes increase approximately in proportion to the change in body length in healthy growing children of average body size
2. Understand methods of measuring pulmonary function in infants, young children, and minimally cooperative children
3. Know that spirometry does not measure absolute lung volumes and capacities

4. Know why slow vital capacity may be greater than forced vital capacity
5. Know the normal interindividual and intraindividual variation in FVC, FEV1, FEF 25-75, and peak expiratory flow

b. Helium dilution
1. Know how to calculate functional residual capacity from helium dilution data
2. Understand the effect of gas trapping or a system leak on helium dilution measurements
3. Understand the principles and limitations of helium dilution measurement of lung volume

c. Nitrogen washout
1. Understand the principle and limitations of nitrogen washout measurements of lung volume
2. Know how to calculate FRC from nitrogen washout data

d. Plethysmography
1. Understand the principles of pressure and volume plethysmography
2. Know that lung cysts and airway obstruction can influence plethysmographic measurements of lung volume
3. Describe why functional residual capacity (FRC) and thoracic gas volume (TGV) may be different when measured by plethysmography versus gas dilution

2. Flows and timed volumes

a. Forced expiratory volumes
1. Know how to calculate forced expiratory flow between 25-75% VC (FEF 25-75) and forced expiratory volume in one second from a sample spirogram
2. Know the reproducibility of PEF, FEF 25-75, FVC, and FEV1 in children
3. Know the criteria for a technically acceptable spirometric trace
4. Know the principle of backward extrapolation for correcting FEV1 measurements

b. Peak expiratory flow
1. Know that peak expiratory flow is effort dependent
2. Understand the limitations of peak expiratory flow rate in evaluating pulmonary function

c. Maximal expiratory flow volume curve
1. Recognize configurations of flow-volume loops that suggest lack of cooperation, poor effort, or glottic closure
2. Know the definitions of flows at various fractions of vital capacity (eg, maximum flow at 50% of vital capacity)
3. Know the variability of flows at various fractions of vital capacity (eg, maximum flow at 50% of vital capacity)
4. Understand criteria for an acceptable flow volume curve, including end of test and reproducibility

3. Diffusing capacity

a. Know that a correction for hemoglobin is required in single-breath diffusing capacity measurements

b. Know that a correction for lung volume is helpful in interpreting single-breath diffusing capacity measurements

c. Know the factors that influence the measurement of single-breath DLCO

4. Airway responsiveness
   a. Bronchodilator testing
      1. Know spirometric criteria for a positive response to bronchodilators
      2. Know that a child with FEV1 that is 100% of predicted value can still show a positive response to bronchodilators
   b. Bronchoprovocation (challenge) testing
      1. Understand the indications for bronchoprovocation testing
      2. Know the time course of bronchoconstriction following different bronchoprovocation challenges (eg, methacholine, cold air, histamine, and antigen challenges)
      3. Know how to interpret a bronchoprovocation test (eg, methacholine, cold air, histamine, and antigen challenges)
      4. Know which medications and foods should be avoided prior to bronchial challenge tests
      5. Know that airway reactivity may fluctuate over time as a function of allergen exposure, respiratory tract infections, and other factors
      6. Know the difference between PC20 and PD20 in bronchial provocation testing
      7. Know the risks of bronchoprovocation testing
      8. Know the contraindications for bronchoprovocation testing
      9. Know how to interpret pulmonary function changes during an exercise test for asthma
5. Exercise testing
   a. Recognize indications for measuring exercise tolerance in children
   b. Know how to recognize the anaerobic threshold on progressive exercise testing
   c. Know that the usual responses to progressive exercise below the anaerobic threshold are linear increases in pulse, respiratory rate, oxygen consumption, and carbon dioxide production
   d. Know that arterial oxygen saturation remains stable and the partial pressure of carbon dioxide in arterial blood remains stable or decreases slightly with exercise in normal children
   e. Know the factors that limit maximum volume of oxygen utilization during exercise
   f. Know the mechanisms that lead to hypoxemia during exercise
   g. Know that above the anaerobic threshold minute ventilation initially increases linearly with carbon dioxide production
6. Respiratory muscle testing
   a. Know how to measure inspiratory and expiratory muscle strength
   b. Recognize indications for measuring inspiratory and expiratory muscle strength in children
   c. Understand limitations of measuring inspiratory and expiratory muscle strength in children
7. Interpretation of pulmonary function tests
   a. Recognize restrictive lung disease on pulmonary function testing
   b. Recognize obstructive lung disease on pulmonary function testing
   c. Recognize the flow-volume loop configurations typical of variable extrathoracic obstruction

d. Recognize the flow-volume loop configurations typical of variable intrathoracic central airway obstruction
e. Recognize the flow-volume loop configuration typical of intrathoracic peripheral airway obstruction
f. Recognize pulmonary function abnormalities associated with inspiratory and expiratory muscle weakness
g. Know that height is the single best predictor of lung function in healthy individuals
h. Know that there are race and sex differences in normal values for pulmonary function tests
i. Recognize the importance of prediction equations in interpreting pulmonary function tests
j. Know the limitations of prediction equations in interpreting pulmonary function test results
k. Recognize the flow-volume loop configuration typical of fixed central airway obstruction
l. Know that in the presence of fixed central airway obstruction spirometry cannot differentiate between intrathoracic and extrathoracic obstruction

8. Pulmonary function testing in infants
   a. Know how to calculate resistance, compliance, and time constant from a passive expiratory flow-volume curve in infants
   b. Know that muscle relaxation is a requirement for passive lung mechanics
   c. Know that passive flow volume curve assesses total respiratory system resistance and compliance in infants
   d. Know the advantages and limitations of partial flow-volume loops in infants
   e. Know how to recognize an obstructive lung defect from a tidal flow-volume curve in infants
   f. Understand the need for sedation in infant pulmonary function testing and the risks involved
   g. Understand the indications for high altitude simulation testing
   h. Understand the methods used in the raised volume thoracic compression technique for measuring airway function in infants
   i. Know the absolute and relative contraindications for measuring pulmonary function in infants using the raised volume thoracic compression technique
   j. Know how to interpret infant spirometry using FEV 0.5, FVC, and forced expiratory flow rates
   k. Understand the methods used in obtaining whole body plethysmography in infants
   l. Know how to interpret infant lung volume measurements using FVC, TLC, and FRC and RV
   m. Know the criteria for a positive bronchodilator response on a flow-volume loop obtained using the raised volume thoracic compression technique in infants
9. Forced oscillometry
   a. Understand the principles and techniques of forced oscillometry for evaluation of respiratory system function
   b. Understand the interpretation, use, and limitations of measurements of respiratory system resistance and reactance using forced oscillometry

c. Understand that forced oscillometry can be used to measure bronchodilator response in infants

D. Invasive procedures
1. Bronchoscopy
a. Flexible vs. rigid
1. Recognize the indications for rigid and for flexible bronchoscopy
b. Indications and risks
1. Recognize complications of bronchoscopy in children
2. Understand the importance of correcting coagulation abnormalities prior to bronchoscopy
3. Know the monitoring requirements for a patient undergoing flexible or rigid bronchoscopy
4. Understand the importance of laryngeal anesthesia prior to bronchoscopic instrumentation
5. Understand the need for, and risks of, sedation in fiberoptic bronchoscopy
c. Anatomic structure-static and dynamic
1. Understand how airway dimensions fluctuate with respiratory efforts during bronchoscopy
2. Understand how sedation affects pharyngeal and laryngeal dynamics
3. Understand how the presence of the bronchoscope in the airway affects airway dynamics
4. Recognize the bronchoscopic appearance of the normal airway and various airway disorders
d. Culture techniques
1. Understand the risks of contamination of specimens of airway secretions obtained by bronchoscopy
2. Know which infectious agents can be cultured from the airways and which can be identified by staining characteristics
e. Bronchoalveolar lavage
1. Know the normal cell population in bronchoalveolar lavage fluid
2. Know the cell population in bronchoalveolar lavage fluid in disease states
3. Recognize the indications for bronchoalveolar lavage in immunocompromised patients
4. Recognize the indications for bronchoalveolar lavage to diagnose noninfectious pulmonary disorders
5. Recognize the complications of bronchoalveolar lavage
f. Transbronchial biopsy
1. Understand the indications for and limitations of transbronchial and endobronchial biopsy
g. Lung biopsy
1. Recognize the histologic appearance of various pathogens on open lung biopsy
2. Understand the indications for and risks associated with open lung biopsy
3. Recognize the histologic appearance of normal lung
4. Understand the indications for and risks associated with thoracoscopic lung biopsy
2. Vascular sampling/access

a. Arterial sampling
    1. Know the indications for and complications of insertion of a pulmonary arterial catheter
    2. Know the techniques for sampling arterial blood
    3. Know the indications for and complications of insertion of a systemic arterial catheter
b. Venous access
    1. Understand the indications for and complications of insertion of central catheters and implantable venous access devices in children with acute and chronic pulmonary disorders

3. Pleural drainage
    a. Recognize the indications for chest tube drainage in children
    b. Know that rapid evacuation of pleural contents can result in re-expansion pulmonary edema or hypotension
    c. Recognize the complications associated with chest tube drainage in children
    d. Understand how a three-chamber system works in conjunction with chest tube drainage
    e. Know the optimal site of placement of a chest tube for the drainage of a pneumothorax or pleural fluid
    f. Understand the indications for video-assisted thoracoscopic techniques for the management of empyema
4. Thoracentesis
    a. Recognize the diagnostic and therapeutic indications for thoracentesis in childhood
    b. Recognize the indications for pleural biopsy in children
    c. Know the importance of proper technique for thoracentesis in minimizing risk of bleeding from intercostal arteries
    d. Know that thoracentesis should be performed in conjunction with ultrasonography or other imaging techniques to drain loculated pleural fluid

E. Imaging
1. X-ray studies
    a. Risks
        1. Know that the amount of radiation exposure associated with x-ray study of the chest is extremely small
    b. Interpretation
        1. Know the ages when the sinuses should be pneumatized and can be visualized on x-ray studies
        2. Recognize the normal positions of the major and minor fissures on x-ray study of the chest
        3. Recognize the radiographic appearance of a normal thymus
        4. Recognize the radiographic appearance of lobar consolidation
        5. Recognize the radiographic appearance of pneumothorax and tension pneumothorax
        6. Recognize the radiographic appearances of paratracheal, mediastinal, and hilar adenopathy
        7. Recognize findings typical of a retropharyngeal abscess on a lateral-view x-ray study of the neck

8. Know which imaging techniques can help distinguish physiologic enlargement from pathologic swelling of the cervical prevertebral space
9. Recognize the radiographic appearance of a right aortic arch
10. Know the differential diagnosis of an opacified hemithorax on radiographic study of the chest
11. Recognize the radiographic appearance of pleural effusion
12. Recognize indications for decubitus x-ray studies
13. Appreciate the difference between anteroposterior-view and posteroanterior-view x-ray studies of the chest
14. Appreciate the difference between upright-view and supine-view radiographic studies of the chest
15. Recognize the radiographic appearance of lobar and segmental atelectasis
16. Recognize the radiographic appearance of malignancy or infection spread by hematogenous contact as opposed to contiguous or airway contact
17. Recognize the significance of air bronchograms on radiographic studies of the chest
18. Recognize the radiographic appearance of pneumomediastinum
19. Recognize the radiographic appearance of bronchiectasis
20. Recognize the radiographic appearance of pneumatocele, lung cyst, and lung abscess
21. Recognize calcification on radiographic study of the chest
22. Recognize the radiographic appearance of normal, decreased, and increased pulmonary blood flow
23. Recognize bony and abdominal abnormalities visible on radiographic studies of the chest
24. Recognize the appearance of bronchial stenosis on radiographic studies
25. Recognize the radiographic appearance of an anterior mediastinal mass
26. Recognize the radiographic appearance of hydatid (echinococcal cysts)
27. Recognize the radiographic appearance of hypertrophic pulmonary osteoarthropathy
28. Recognize the radiographic appearance of a normal lung
29. Recognize the radiographic abnormalities that may occur with an inhaled foreign body
30. Recognize miliary shadowing on a radiographic study

2. Fluoroscopy
   a. Risks
      1. Understand that fluoroscopy delivers a larger dose of radiation than standard radiography
   b. Indications
      1. Understand indications for chest fluoroscopy
   c. Interpretation
      1. Understand the typical fluoroscopic appearance of unilateral and bilateral diaphragm paralysis
   d. Esophagography/swallowing cine-esophagography
      1. Describe the use and limitations of esophagography in defining mediastinal anatomy

3. Angiography
    a. Know the indications for pulmonary and bronchial arteriography
    b. Indications
        1. Know the indications for pulmonary and bronchial angiography
    c. Interpretation
        1. Recognize arteriovenous malformation on pulmonary angiography
        2. Recognize V/Q scan appearance in pneumonia or atelectasis
4. Computed tomography (CT scan)
    a. Know that computed tomography (CT scan) of the chest provides definition of mediastinal anatomy and intrathoracic masses
    b. Know that high-resolution thoracic computed tomography (CT scan) is more sensitive than standard radiography in assessing bronchiectasis and interstitial processes
    c. Know that contrast can be used to differentiate vascular from nonvascular masses on computed tomography (CT scan)
    d. Know the indication for helical chest computed tomography
    e. Recognize that chest computed tomography delivers more radiation than standard x-ray studies
5. Magnetic resonance imaging (MRI)
    a. Know that magnetic resonance imaging (MRI) provides more detailed definition of central vascular structures than computed tomography (CT scan)
    b. Understand the difficulties of monitoring children during magnetic resonance imaging (MRI)
    c. Know that magnetic resonance imaging (MRI) does not provide definition of bones or lung tissue
    d. Be able to interpret findings on magnetic resonance imaging of the chest
6. Ultrasonography
    a. Recognize the indications for ultrasonography of the chest
    b. Recognize the role of ultrasonography in the evaluation of pleural effusions
7. Nuclear medicine
    a. Recognize the indications for ventilation- perfusion lung scan
    b. Be able to interpret the findings on ventilation-perfusion lung scan

F. Laboratory diagnostic studies
1. Sputum/nasopharyngeal washes
    a. Know the characteristics of an adequate sputum sample
    b. Recognize alveolar macrophages, ciliated cells, leukocytes, and squamous epithelial cells in sputum
    c. Know that the presence of squamous epithelial cells indicates oropharyngeal contamination of sputum samples
    d. Know that bacterial culture of sputum samples is complicated by oropharyngeal contamination
    e. Understand the method of and rationale for obtaining gastric aspirates for mycobacterial culture
    f. Know that cytology can be used to identify cytomegalovirus infection
2. Pleural fluid analysis (see pleural diseases)
3. Blood analysis (see respiratory failure)

4. Oximetry (see therapy)
5. Biopsy techniques
    a. Recognize indications for lung biopsy
    b. Understand the difference in histologic appearance between caseating and noncaseating granulomas in the lung
    c. Recognize indications for applying special stains to biopsy material (eg, silver stains, immunofluorescence, fungal stains)
    d. Recognize the indications for electron microscopy of biopsy specimens of pulmonary tissue
6. Molecular biologic approaches
    a. Know the role of PCR techniques in the diagnosis of pulmonary diseases
7. Exhaled nitric oxide
    a. Recognize the techniques, indications, and limitations of exhaled nitric oxide measurement

3. **Therapy**

A. Pharmacologic principles
    1. Volume of distribution of drugs
        a. Understand the definition and principles of volume of distribution of a drug
        b. Know that volume of distribution of drugs varies throughout infancy and childhood due to changes in body composition
    2. Clearance/serum half-life
        a. Know that if a drug is administered at intervals equivalent to its half-life that it will take approximately five half-lives to reach 97% of the final steady-state plasma concentration
        b. Know the importance of a loading dose of drugs to achieve therapeutic concentrations rapidly
    3. Concentration of drugs in lung secretions/serum
        a. Understand the relationship between serum concentrations and sputum concentrations of drugs administered systemically

B. Bronchodilators (see asthma)
    1. Adrenergic drugs
        a. Indications
            1. Know the indications for the administration of epinephrine during resuscitation
            2. Recognize the indications for the use of alternate routes of administration to deliver adrenergic agents (eg, ET tube)
        b. Mechanism of action
            1. Understand the mechanism of action of the alpha and beta adrenergic agents
        c. Clinical effects
            1. Recognize actions of alpha and beta adrenergic drugs other than those on smooth muscle
            2. Know the relative beta-2-adrenergic receptor selectivity of adrenergic drugs
            3. Know the various routes available for delivery of beta-2 drugs and the indications for their use
        d. Duration of action
            1. Know the duration of action of adrenergic drugs that are commonly administered

e. Toxicity and side effects
    1. Recognize the side effects of the various adrenergic agents
    2. Recognize the side effects of beta-2 drugs administered by inhalation, as opposed to orally
    3. Recognize the potential risks of long-acting beta-2 agonists
2. Theophylline
    a. Indications
        1. Know that theophylline may be indicated for symptoms of nocturnal asthma and is an alternative, but not preferred, long-term preventative asthma therapy if cost or compliance with inhaled medications is considered
        2. Know that theophylline bolus and drip are only infrequently indicated in status asthmaticus and then only in the most severe cases
    b. Mechanism of action
        1. Know the proposed mechanism of action of theophylline
    c. Clinical effects
        1. Recognize nonbronchodilating effects of theophylline
        2. Know when blood theophylline concentrations should be measured: acute therapy, chronic therapy
    d. Duration of action
        1. Recognize drugs or conditions that affect clearance of theophylline
    e. Toxicity and side effects
        1. Recognize the signs of acute theophylline overdose
        2. Know the treatment of acute theophylline overdose
        3. Recognize the chronic side effects of theophylline therapy
3. Anticholinergics
    a. Indications
        1. Recognize indications for administration of anticholinergic drugs to patients with lung disease
    b. Mechanism of action
        1. Understand the mechanism of action of ipratropium and tiotropium bromide
    c. Clinical effects
        1. Understand bronchodilating and nonbronchodilating actions of anticholinergic drugs
    d. Duration of action
        1. Anticipate the time of peak response and the expected duration of action of anticholinergic drugs
    e. Toxicity and side effects
        1. Recognize the side effects of ipratropium and tiotropium
        2. Recognize the side effects of atropine
        3. Know why ipratropium has fewer side effects than atropine

C. Corticosteroids
1. Indications
    a. Know when corticosteroids should be added to the therapeutic regimen of a patient
2. Mechanism of action

a. Know that corticosteroids have a broad action on the inflammatory process, including suppression of cytokine generation, airway eosinophil recruitment, and inflammatory mediator release

3. Clinical effects
   a. Know that corticosteroids can restore beta-adrenergic responsiveness, block the late reaction to allergen, and reduce airway hyper-responsiveness
4. Duration of action
   a. Understand the relative differences in duration of action of common corticosteroid preparations
5. Toxicity and side effects
   a. Recognize the clinical side effects of corticosteroids administered by inhalation or systemically
   b. Know that cataracts can result from treatment with systemic corticosteroids
   c. Know the impact of corticosteroids on growth
   d. Know that chronic immune suppression may follow corticosteroid therapy in children
   e. Know that long-term corticosteroid therapy may be associated with corticosteroid-induced myopathy

D. Cromolyn sodium and nedocromil
1. Indications
2. Mechanism of action
3. Clinical effects
   a. Know that neither cromolyn sodium nor nedocromil acts as a bronchodilator but will block bronchospasm induced by allergen challenge, exercise, and cold air
4. Duration of action
5. Toxicity and side effects

E. Leukotriene modifiers
1. Indications
   a. Know the indications for leukotriene modifiers in the management of asthma
2. Mechanism of action
   a. Know the mechanisms of action for the different types of leukotriene modifiers
3. Clinical effects
   a. Recognize the effectiveness of leukotriene modifiers compared to other asthma-controlling medications
   b. Understand the role of leukotriene modifiers in the management of nocturnal and exercise-induced asthma
4. Duration of action
   a. Know the duration of action of leukotriene modifiers
5. Toxicity and side effects
   a. Understand that side effects of leukotriene modifiers are infrequent

F. Diuretics
1. Indications (see specific diseases)
2. Mechanism of action
   a. Understand the renal and nonrenal actions of diuretics
3. Clinical effects
4. Duration of action

5. Toxicity
    a. Understand how diuretic therapy may contribute to the development of metabolic alkalosis
    b. Know the interaction of diuretics and aminoglycosides with respect to eighth nerve toxicity
    c. Understand the toxicity of chronic diuretic therapy in infants and children

G. Chest physiotherapy
1. Indications
    a. Know that chest physiotherapy is indicated in the treatment of chronic, suppurative lung disease
    b. Know that chest physiotherapy is not indicated in the treatment of uncomplicated pneumonia
2. Techniques
    a. Know the various techniques for performing chest physiotherapy (eg, autogenic drainage techniques, positive expiratory pressure, mask, flutter device, high-frequency chest wall oscillations, intrapulmonary percussive devices, etc.)
    b. Know the role of cough-assist devices in patients with muscular weakness

H. Aerosol therapy
1. Factors determining deposition site in the airway
    a. Understand the relationship between particle size and deposition site in the airway
    b. Know that gravity, inertia, and Brownian movement are mechanisms contributing to deposition of an aerosol in the airway
2. Types of equipment
    a. Nebulizer
        1. Understand the differences between ultrasonic and jet nebulizers
        2. Recognize that in aerosol therapy the majority of medication is not delivered to the lung
    b. Metered-dose inhaler
        1. Know the techniques for maximizing the response of a patient to medication administered by metered-dose inhaler
        2. Understand the value of spacer devices in metered-dose inhalers

I. Humidification
1. Understand the difference between relative humidity and absolute water content
2. Know techniques for humidifying inspired gas and appreciate the relative efficiency of each
3. Recognize the indications for humidification of inspired gases
4. Recognize the side effects of mist therapy

J. Oxygen therapy
1. Delivery systems
    a. Recognize the differences among various oxygen delivery systems (eg, liquid, compressed gas, concentrator)
    b. Know that when nasal prongs are used, during the process of inspiration there is a variable and unknown inspired oxygen concentration
    c. Know the underlying principle of a Venturi valve
    d. Know which oxygen delivery systems provide a known fraction of inspired oxygen
    e. Know uses and risks of hyperbaric oxygen therapy for carbon monoxide poisoning

f. Understand the relative advantages and disadvantages of oxygen delivery via nasal prongs, face mask, blow by, and tent
g. Know that the gas flow at the end of a length of tubing attached to an oxygen source is independent of the length of the tubing
h. Know the limits of using an oxygen concentrator (ie, high-flow)

2. Indications
   a. Recognize clinical indications for oxygen therapy
   b. Know the differences between oxygen saturation, oxygen content, and oxygen partial pressure
   c. Recognize the cyanotic conditions that will not respond to oxygen therapy
   d. Recognize the indications for oxygen therapy during air travel in patents with lung disease
3. Monitoring systemic oxygenation
   a. Know the principles underlying pulse oximetry
   b. Know factors that influence the accuracy of pulse oximetry
   c. Know technical factors that may alter the reading of oxygen saturation (eg, fetal hemoglobin, carbon monoxide, methemoglobin)
   d. Understand the usefulness and limitations of determination of the mixed venous oxygen content
   e. Know that oximeters require adequate tissue perfusion/blood flow before oxygen saturation can be accurately assessed
   f. Understand the limitations of transcutaneous PO2 electrodes
   g. Understand the effect of increased PaO2 on mixed venous PO2
   h. Understand the conditions under which capillary PO2 does not accurately reflect arterial PO2
4. Toxicity/side effects of oxygen therapy
   a. Know the appropriate amount of oxygen to administer to a patient in chronic respiratory failure
   b. Recognize the adverse effects of oxygen on the various pulmonary cells
   c. Understand the major cellular antioxidant systems
   d. Understand the effect of fetal lung maturation or previous oxygen exposure on cellular antioxidant enzyme systems
   e. Recognize the clinical sequence of symptoms due to oxygen toxicity
   f. Know that it is the arterial PO2 that is important in the pathogenesis of retinopathy of prematurity, whereas it is the alveolar PO2 that is important in the pathogenesis of lung oxygen toxicity
   g. Know that the time of onset of oxygen toxicity is proportional to the alveolar PO2 (eg, normobaric versus hyperbaric oxygen)
   h. Know the mechanism by which oxygen promotes atelectasis
5. Strategies to prevent oxygen toxicity

K. Mechanical ventilation
1. Indications for use
   a. Recognize the indications for mechanical ventilation in various clinical situations
   b. Recognize the indications for discontinuing mechanical ventilation in various clinical situations
   c. Know approaches to weaning from mechanical ventilation

2. Modes
    a. Understand the difference between jet and high- frequency oscillatory ventilation
    b. Know the definitions of intermittent mandatory ventilation, synchronous intermittent mandatory ventilation, and pressure support
    c. Understand the principles and use of intermittent mandatory ventilation; pressure support; proportional-assist, assist-control, and controlled ventilation; and airway pressure release ventilation
    d. Recognize the indications for use of high-frequency oscillation and jet ventilation
    e. Know the limitations of high-frequency oscillation and jet ventilation
    f. Understand the principles of and indications for time-cycled ventilation
    g. Recognize the clinical indications for intermittent mandatory ventilation, pressure support, assist- control ventilation, and controlled ventilation
    h. Recognize the indications for use of negative pressure ventilation
3. Volume- vs pressure-limited mechanical ventilation
    a. Know the definition of volume-limited ventilation
    b. Know the definition of pressure-limited ventilation
    c. Understand the principles of pressure-limited ventilation and volume-limited ventilation and recognize clinical indications for their use
4. Ventilatory parameters
    a. Inspiratory/expiratory time
        1. Know the definition of inspiratory time
        2. Understand the effect of inspiratory time on gas distribution
        3. Understand the relationship between inspiratory time and peak pressure
        4. Understand the relationship between inspiratory time and peak flow
        5. Understand the importance of expiratory time with respect to gas trapping during mechanical ventilation
        6. Know how to change inspiratory time to accommodate various clinical situations
    b. Tidal volume
        1. Recognize the factors that affect tidal volume delivered to a patient by a mechanical ventilator
        2. Understand the relationship between tidal volume and airway pressure in various modes of ventilation
        3. Quantitate the effects of compressible volume on delivered tidal volume
        4. Know how to choose an appropriate ventilator tidal volume in various clinical settings
    c. Rate
        1. Know how to choose an appropriate ventilator respiratory rate in various clinical situations
    d. Wave form
        1. Understand the relationship between inspiratory wave form and mean airway pressure
        2. Understand the differences in flow-wave form in pressure-limited and volume-limited ventilatory modes
    e. I:E ratio

1. Appreciate the potential effects of altering the I:E ratio on arterial blood gas values in various disease states
2. Appreciate the potential impact of altering the I:E ratio on air trapping
3. Be able to calculate inspiratory time from rate and I:E ratio during mechanical ventilation

f. PEEP and CPAP
1. Understand the mechanisms of action of positive end-expiratory pressure
2. Understand the physiologic effects of positive end-expiratory pressure
3. Understand the mechanisms of the side effects of positive end-expiratory pressure
4. Know the methods available for the application of positive end-expiratory pressure to patients who are receiving ventilation, as well as to those who are not

5. Home mechanical ventilation
a. Chronic respiratory failure
1. Understand that ventilatory muscle power must be sufficient to overcome the respiratory load in chronic respiratory failure
2. Know that respiratory failure occurs when normal ventilatory muscles cannot overcome increased respiratory loads and/or weak ventilatory muscles cannot overcome normal respiratory load
3. Understand that central respiratory drive must be sufficient to overcome the respiratory load in chronic respiratory failure
4. Know that combined ventilatory muscle weakness and central drive make respiratory failure more likely

b. Initiating home mechanical ventilation
1. Know that home mechanical ventilation is best initiated electively when the need can be predicted, such as in neuromuscular disease patients with CO2 retention shown on polysomnography
2. Know that candidates for home mechanical ventilation must have a stable respiratory disorder that does not require frequent changes in ventilator settings
3. Know that candidates for home mechanical ventilation include children who cannot be weaned from assisted ventilation following consistent weaning attempts when the child is otherwise stable

c. Positive pressure ventilation via tracheostomy
1. Know that positive pressure ventilation via a tracheostomy is the most common method of home mechanical ventilation and that it can be used in children requiring full-time or part-time assisted ventilation
2. Know that small, uncuffed tracheostomy tubes are preferred to prevent tracheomalacia and to permit speech in patients undergoing positive pressure ventilation via tracheostomy tube
3. Know that small, uncuffed tracheostomy tubes have large variable leaks that must be compensated for using ventilator techniques in patients receiving positive pressure ventilation via tracheostomy tube
4. Know that ventilators are often used in a pressure-control or time-cycled pressure-limited mode to compensate for variable leaks around uncuffed

tracheostomy tubes in patients receiving positive pressure ventilation via tracheostomy tube

5. Know that mucous plugging of tracheostomies is common, and patients should be monitored with appropriate alarm systems to detect tracheostomy plugging and/or decannulation during positive pressure ventilation via the tracheostomy
6. Know that high pressure alarms are used to detect tracheostomy plugs but that they will not sound if a ventilator is used in a pressure control mode during positive pressure ventilation via tracheostomy
7. Know that low pressure alarms are used to detect a ventilator circuit disconnect or a tracheostomy decannulation during positive pressure ventilation via tracheostomy tube

d. Bi-level positive pressure ventilation by mask or nasal prongs
   1. Know that during bi-level positive airway pressure ventilation by mask or nasal prongs baseline expiratory positive pressure (E-PAP) with inspiratory positive airway pressure (I-PAP) are delivered to assist a child's spontaneous breathing
   2. Know that bi-level positive pressure ventilation is delivered via a nasal mask, facemask, or nasal prongs, so a tracheostomy is not required
   3. Know that the different modes of bi-level positive pressure ventilation include spontaneous, timed, spontaneous/timed, and continuous positive airway pressure (CPAP)
   4. Know that during bi-level positive pressure ventilation tidal volume is proportional to the I-PAP minus E-PAP difference
   5. Know that bi-level positive pressure ventilation is most commonly used for children requiring ventilation only at night
   6. Know that the complications of bi-level positive pressure ventilation are most frequently related to the facemask
e. Negative pressure ventilation
   1. Know that during negative pressure ventilation, ventilation is caused by negative inspiratory pressure applied to the outside of the chest and upper abdomen, so a tracheostomy is not required
   2. Know that during negative pressure ventilation adequate ventilation depends on the ability to expand the chest with negative pressure applied outside the chest, making negative pressure ventilation less effective when chest wall motion is restricted
   3. Know that upper airway obstruction can occur with negative pressure ventilation
f. Diaphragm pacing
   1. Know that diaphragm pacing requires surgical implantation of phrenic nerve electrodes and receivers
   2. Know that diaphragm pacing is contraindicated in patients with damage to the phrenic nerve (motor neuropathies or trauma) or primary myopathy of the diaphragm
   3. Know that diaphragm pacing is most commonly used for children with congenital central hypoventilation syndrome, other central hypoventilation syndromes, and high spinal cord injury (C1-2)

4. Know that for full-time ventilator-dependent patients, diaphragm pacing can be used during wakefulness to improve mobility and rehabilitative potential
5. Know that for patients who are ventilator-dependent only during sleep, diaphragm pacing may be used to remove a tracheostomy, but upper airway obstruction may occur during diaphragm pacing in a sleeping child without a tracheostomy
6. Know that the complications of diaphragm pacing include phrenic nerve injury during surgery, infection of an implanted foreign body, and unknown long-term sequelae of phrenic nerve stimulation

6. Risks/complications
   a. Recognize the risks and complications of mechanical ventilation
7. Intubation
   a. Recognize the complications associated with endotracheal intubation in children
   b. Recognize the indications for selective bronchial intubation
   c. Recognize the indications for endotracheal intubation in children

L. Home monitoring
1. Recognize the indications for home monitoring
2. Understand that the benefit of home monitoring in the prevention of sudden infant death syndrome is unproven

M. Incentive spirometry
1. Recognize the indications for incentive spirometry

N. Adherence
1. Know the methods of assessing drug adherence in patients with respiratory disorders
2. Know that lack of adherence is a major factor in failure rates of prescribed therapeutic regimens
3. Know ways to enhance patient adherence with therapeutic regimens
4. Recognize factors that contribute to poor adherence with therapy

O. Lung transplantation
1. Indications
   a. Recognize clinical indications for heart-lung, bilateral lung, and single-lung transplantation
2. Complications
   a. Know that most acute rejection episodes following heart-lung or lung transplantation occur in the first three months after the surgery
   b. Recognize the signs and symptoms of acute rejection following heart-lung or lung transplantation
   c. Know that transbronchial biopsy is the standard method for diagnosing acute rejection following heart-lung or lung transplantation
   d. Recognize the usual histologic pattern of acute rejection following heart-lung or lung transplantation
   e. Know that obliterative bronchiolitis is the most frequent pattern of chronic rejection following heart-lung or lung transplantation
   f. Recognize the common agents that cause pulmonary infections following heart-lung or lung transplantation
   g. Understand the role of bronchoalveolar lavage in establishing the etiology of pulmonary infection following heart-lung or lung transplantation

h. Understand the importance of bronchial blood flow to the trachea and bronchi in transplanted lungs
i. Know the complications of lung transplantation
j. Understand that the bronchial airways are denervated following heart-lung or lung transplantation

P. Tracheostomy
1. Indications
a. Recognize the clinical indications for tracheostomy in children
2. Complications
a. Recognize the immediate postoperative complications of tracheostomy
b. Implement appropriate therapeutic steps when a child with a tracheostomy suddenly becomes cyanotic or has other signs and symptoms of acute hypoventilation
c. Recognize the chronic complications of tracheostomy
d. Recognize that a child younger than 1 year of age who has a tracheostomy because of severe subglottic stenosis is at risk for increased mortality during the first year after birth
e. Recognize that aspiration secondary to dysfunctional swallowing may complicate tracheostomy
f. Know the indications and contraindications of speaking valves in children with tracheostomy
3. Clinical effects
a. Know how to confirm airway adequacy prior to decannulation in a child with a tracheostomy

Q. Dornase alfa
1. Indications
a. Understand the indications for the use of nebulized dornase alfa
2. Mechanism of action
a. Know the mechanism of action of dornase alfa
3. Clinical effects
a. Know that dornase alfa improves lung function in patients with cystic fibrosis and mild to moderate lung disease
4. Toxicity and side effects
a. Know the common side effects of dornase alfa

R. Miscellaneous
1. Understand that dopamine increases peripheral vascular resistance and that dobutamine decreases peripheral vascular resistance
2. Know the medications that increase respiratory drive
3. Know that nitric oxide is a potent vasodilator
4. Know that nitric oxide modulates interactions between inflammatory cells and vascular epithelium
5. Know that nitric oxide may improve ventilation/perfusion mismatch and reduce pulmonary artery pressure
6. Know that nitric oxide use requires accurate measurement of NO concentration, careful titration to the lowest effective concentration, and gradual termination

4. **Prevention**

A. Immunizations
    1. Pertussis
        a. Composition
        b. Efficacy
            1. Know that it takes a series of pertussis vaccinations to achieve protection in young children
            2. Know the efficacy of acellular pertussis vaccination
            3. Know that a booster (Tdap) is recommended for adolescents 11 to 18 years of age
            4. Understand the concept of herd immunity and the increased risk for pertussis in the absence of widespread immunity in a population
        c. Complications
            1. Know the safety of the acellular pertussis vaccination
    2. Diphtheria
        a. Composition
            1. Know that diphtheria vaccine is a toxoid
        b. Efficacy
            1. Know that a series of diphtheria vaccinations is required to achieve protection
    3. Haemophilus influenzae
        a. Composition
        b. Efficacy
            1. Know the efficacy of Haemophilus influenzae vaccine for respiratory disease
    4. Measles
        a. Composition
            1. Know that the measles vaccine is a live virus preparation
        b. Efficacy
            1. Know that two doses of measles vaccine are recommended for normal children
        c. Complications
            1. Recognize the absolute contraindications for measles vaccination
    5. Tuberculosis
        a. Composition
            1. Know that bacille Calmette-Guerin vaccine consists of a live attenuated strain of Mycobacterium bovis
        b. Efficacy
            1. Recognize the indications for bacille Calmette Guerin vaccine in developed and undeveloped countries
            2. Know the efficacy of bacille Calmette Guerin vaccine
        c. Complications
            1. Know how to interpret results of a tuberculin skin test in a patient who has been vaccinated with bacille Calmette Guerin vaccine
            2. Recognize the complications of bacille Calmette Guerin vaccine
    6. Pneumococcal
        a. Composition
            1. Know that 23-valent pneumococcal vaccine is composed of purified capsular polysaccharide from the serotypes that most commonly cause serious pneumococcal disease

2. Know that heptavalent pneumococcal vaccine is composed of a protein conjugate vaccine derived from serotypes most likely to cause invasive disease in young children (i.e., meningitis and bacteremic pneumonia)
3. Understand the differences between pediatric heptavalent conjugate pneumococcal vaccine and the 23-valent polysaccharide vaccine in indication and duration of protection

b. Efficacy
1. Recognize the clinical conditions for which pneumococcal vaccine is indicated
2. Know that pneumococcal vaccine does not confer absolute protection to children at high risk
3. Know the immunologic response to immunization

7. Influenza
a. Composition
1. Recognize that influenza vaccine is available in two forms: formalin inactivated vaccine and a live attenuated vaccine

b. Efficacy
1. Recognize the clinical indications for administration of influenza vaccine
2. Recognize the limitations of influenza vaccine
3. Know the recommendations for administration of inactivated and live attenuated influenza vaccine
4. Know that prophylactic oseltamivir therapy is effective against influenza A virus
5. Know that short-term administration of systemic corticosteroids does not suppress the protective effect of influenza vaccine
6. Know that live attenuated influenza vaccine is contraindicated in children with asthma

B. Smoking
1. Active smoking
a. Demographics of onset
1. Know that the great majority of adult smokers are already committed smokers by the time they are 18 years of age

b. Factors influencing onset
1. Know that peer pressure is the primary influence on smoking initiation

c. Pulmonary and extrapulmonary health effects
1. Recognize that smoking is the number one preventable cause of death in the United States
2. Know the factors that influence the effectiveness of smoking cessation programs

2. Passive smoking - health effects
a. Fetus
1. Know that maternal smoking leads to an increase in fetal loss, a reduction in birth weight, and an adverse impact on lung growth and function

b. Infant
1. Know that passive smoking leads to increased rates of both upper and lower respiratory tract infections and sudden infant death syndrome in infants

c. Child

1. Know that passive smoking may be a potent stimulus for bronchoconstriction in children with pre-existing reactive airways disease

3. Interventions
   a. Preventions
      1. Know that smoking prevention would be the most effective public health measure possible in terms of reducing mortality in the United States
   b. Smoking cessation
      1. Know that effective smoking cessation must take into account the fact that for many people smoking is both a habit and an addiction

C. Neonatal infections
   1. Know that treatment of pregnant women colonized with Chlamydia trachomatis may prevent disease in the infant
   2. Know that routine eye prophylaxis in a newborn infant colonized with Chlamydia trachomatis does not prevent the onset of pneumonia

5. **Lung Growth and Development**

A. Fetal
   1. Structure
      a. Embryonic stage
         1. Know that the lung arises as an outpouching of the primitive gut
         2. Know the characteristics of the embryonic stage of lung development
         3. Know that the major bronchial divisions are formed by 16 weeks of gestation
         4. Know that transcription factors direct branching morphogenesis
         5. Know that transcription factors are involved in cell commitment and differentiation
         6. Know the postconceptional time of the embryonic stage (i.e., 3 to 6 weeks postconception)
         7. Know that normal airway branching involves signaling between endoderm and mesoderm and that the mesoderm directs the branching pattern
      b. Pseudoglandular stage
         1. Know the histologic characteristics of the pseudoglandular stage of lung development
         2. Know the postconceptional time of the pseudoglandular stage (i.e., 6 to 16 weeks postconception)
         3. Know that most gross congenital abnormalities in lung development (bronchopulmonary sequestration, cystic adenomatoid malformation, and congenital diaphragmatic hernia) arise during the pseudoglandular phase of development
      c. Canalicular stage
         1. Know the histologic characteristics of the canalicular stage of lung development
         2. Know the postconceptional time of the canalicular stage (i.e., 16 to 26 weeks postconception)
      d. Saccular stage
         1. Know the histologic characteristics of the saccular stage of lung development
         2. Know the postconceptional time of the saccular stage (i.e., 26 to 36 weeks postconception)
      e. Alveolar stage

1. Know the histologic characteristics of the alveolar stage of lung development
2. Know the postconceptional time of the alveolar stage (i.e., 36 weeks postconception through early childhood)

f. Cellular structure
1. Know which cellular characteristics of the airway vary with fetal lung development
2. Know which cellular characteristics of the airspace vary with fetal lung development
3. Know which cellular characteristics of the pulmonary vasculature vary with fetal lung development

2. Physiology/Pathophysiology

a. Circulation
1. Know that the fetal pulmonary circulation receives only minimal amounts of the combined ventricular output (approximately 5% to 7%)
2. Know that the systemic circulation has a low resistance circulation due to the placental vascular bed
3. Understand the mechanisms that maintain a high pulmonary vascular resistance in utero

b. Airway and alveolus
1. Understand the mechanisms of production and the unique composition of fetal lung liquid
2. Know that normal lung development is dependent upon normal secretion of lung liquid
3. Understand the developmental timing and regulation of surfactant synthesis
4. Know the factors that interfere with alveolar development (external and internal limitation of space, oligohydramnios, fetal breathing abnormalities, maternal smoking)
5. Understand the role of surfactant proteins in surfactant homeostasis
6. Know the genetic defects associated with congenital pulmonary alveolar proteinosis
7. Know that obstruction to outflow of fetal lung liquid (e.g., laryngeal atresia) results in pulmonary hyperplasia

c. Fetal breathing
1. Recognize the pattern of fetal breathing
2. Know that fetal breathing pattern is influenced by sleep state
3. Know that fetal breathing influences lung development

d. Regulation
1. Understand the influence of growth factors on lung development
2. Understand the effects of thyroid hormones on lung development
3. Understand the effects of corticosteroid hormones on lung development
4. Know that mesenchymal cells play a directive role in the development of epithelium through a paracrine/autocrine interaction with the epithelium

B. Perinatal

1. Circulation

a. Know the mechanical, chemical, and vasoactive signals involved in lowering pulmonary vascular resistance after birth

b. Know the role of nitric oxide in lowering pulmonary vascular resistance

2. Airflow and lung mechanics
   a. Understand how increased surface tension at the alveolar air-liquid interface can lead to reductions in end-expiratory volume
   b. Understand the role of laryngeal and respiratory muscle function in maintaining end-expiratory volume above functional residual capacity in neonates and young infants
   c. Know that surfactant is released from type II cells at birth
3. Gas exchange
   a. Know the stimuli involved in initiation of respiration (thermal, tactile, chemical, etc.) after birth
   b. Know changes in arterial blood gas tensions and acid base status during the perinatal period
4. Clearance of lung liquid
   a. Know that lung liquid clearance takes many hours
   b. Know pathways of lung liquid removal during and following the birth process
   c. Understand the mechanisms of lung liquid removal (mechanical, osmotic forces, active sodium transport) during and following the birth process

C. Postnatal
1. Structure
   a. Airways and alveoli
      1. Know that continuing alveolarization of terminal bronchioles “transforms" them into respiratory bronchioles
      2. Know that new alveoli arise from saccules by the processes of septation and capillary invasion
      3. Know that collateral ventilation is poorly developed in newborn infants
      4. Know the timing of the greatest increases in alveolar numbers during postnatal life
      5. Know that there is no increase in the number of small conducting airways after birth
      6. Know that vascular endothelial growth factor is important in lung vasculogenesis
   b. Pulmonary circulation
      1. Understand the association between decreased pulmonary arterial pressure and decreased muscularization of the pulmonary circulation
      2. Know that proliferation of new arteries occurs most rapidly in the first one to two years after birth and parallels alveolar multiplication
      3. Understand the structural changes of the pulmonary arterial system that occur postnatally
      4. Know that there are nonmuscularized pulmonary arteries
      5. Know that pulmonary veins increase in number postnatally in association with alveolar multiplication
   c. Cellular growth
      1. Know that neuroepithelial bodies or argyrophil cells are relatively prominent in the lungs of newborn infants, decrease in density during the first year after birth, and increase in infants with bronchopulmonary dysplasia

2. Physiology/pathophysiology
    a. Understand the relation between lung growth and diffusing capacity for carbon monoxide
    b. Understand the effect of restriction of the chest wall on growth of the lung
    c. Understand the effect of living at a high altitude on lung growth
    d. Understand the effect of pneumonectomy on the growth of the remaining lung and thoracic cage
    e. Know that lung distention influences growth of the remaining lung after pneumonectomy
    f. Know the roles of surfactant proteins in host defenses
    g. Understand the impact on lung growth of reduced inspiratory force in myopathic patients

6. **Structure and Function of the Respiratory System**

A. Upper airway

1. Nose
    a. Know the anatomy of the nasal turbinates and the sinus ostia
    b. Understand the function of the turbinates in terms of increasing surface area in the nose
    c. Understand the function of the nasal passage in terms of humidification and warming of the inspired air
    d. Know that the nose accounts for approximately half of total inspiratory airway resistance
    e. Know that posture affects nasal patency, ie, resistance is increased when patients are in the supine position
    f. Know that nasal airflow resistance decreases during exercise
    g. Understand the importance of the muscles of the upper airway (including the genioglossus, alar nasal and laryngeal abductors) in maintaining airway patency during inspiration
    h. Know the cellular components of the nasal epithelium
2. Pharynx
    a. Nasopharynx/choanae
        1. Know the location of the adenoids
        2. Understand the importance of velopharyngeal closure in speech and swallowing
    b. Oropharynx
        1. Understand the steps in normal swallowing (closure of the glottis and velopharyngeal valve, coupled with cricopharyngeal sphincter relaxation)
        2. Understand the function of the genioglossus in determining airway patency
3. Larynx
    a. Know the normal anatomy of the larynx and the supralaryngeal areas
    b. Understand the normal function of the vocal cords in protecting the airway
    c. Know the relative change in position of the larynx during growth in the neonate as compared to in the adult (C3-4 versus C5)
    d. Understand the innervation of the larynx
    e. Know that the cricoarytenoid posterior is the only muscle capable of opening the larynx
4. Subglottic space

a. Know that the cross-sectional area at the cricoid is fixed and not affected by transmural pressure changes
b. Know that below the cricoid, the tracheal rings are C-shaped and the cross-sectional area is variable with changes in transmural pressure
c. Know that compliance of the trachea is greater in children than in adults

B. Thorax
1. Skeleton
a. Know the normal anatomy of the rib cage
b. Understand the changes in the rib cage that are associated with respiration
c. Understand the physiologic significance of maturational ossification of the rib cage
2. Respiratory muscles
a. Diaphragm
1. Know that the mechanical advantage of the diaphragm depends on its domed shape
2. Know that the diaphragmatic muscle has two components: a sternal (costal) portion and a crural portion
3. Understand the relationship between diaphragmatic fatigue and changes in chest and abdominal wall motion
4. Know that the diaphragm is innervated by the phrenic nerve (C3, C4, and C5)
5. Know the course of the phrenic nerve from the neck to the diaphragm
6. Know that the diaphragm is the primary muscle of respiration during quiet breathing
7. Know that the diaphragm is composed of different fiber subtypes
8. Know that inspiratory muscles may remain active during expiration in some lung diseases
9. Identify the anatomic attachments of the diaphragm
10. Understand the functional significance of the costal and crural regions of the diaphragm
11. Understand the embryologic development of the diaphragm
12. Understand the length-tension relationship of the diaphragm
13. Understand how the length-tension relationship influences diaphragmatic function in hyperinflated lungs
14. Understand how diaphragmatic contraction results in inspiration
15. Know that quiet expiration is a passive process, ie, there is no active diaphragmatic contraction
16. Know that in infants the diaphragm attachment to the lower rib cage is more perpendicular than in children and adults
17. Understand that intercostal muscle weakness may contribute to the development of scoliosis
b. Intercostal muscles
1. Know that intercostal muscles are important in forced expiration and cough
2. Know that intercostal muscles contract synchronously with the diaphragm to maintain rib cage stability
c. Accessory muscles
1. Know that the sternocleidomastoid muscles are used only during increased work of breathing and are not active at rest

2. Understand how the scalene and sternocleidomastoid muscles contribute to respiration

d. Abdominal
1. Recognize the role of abdominal muscles in respiration, ie, stabilizing chest wall and expiratory movements (cough and exercise)
2. Understand the role of the abdominal muscles in cough

3. Neurovascular supply
a. Know the location of the neurovascular bundle supply to the intercostal muscles

4. Pleural space (see pleural diseases)
a. Parietal pleura
1. Understand the contribution of the parietal pleura to the production and clearance of pleural fluid
2. Understand the innervation of the parietal pleura

b. Visceral pleura
1. Understand the innervation of the visceral pleura
2. Understand the contribution of the visceral pleura to the production and clearance of pleural fluid

5. Mediastinum
a. Recognize the boundaries of the various compartments of the mediastinum (anterior, posterior, superior, inferior)
b. Recognize the normal structures located in each of the four mediastinal compartments (anterior, posterior, superior, inferior)
c. Recognize the abnormal structures that may be located in each of the four mediastinal compartments

C. Lower airway
1. Trachea
a. Length, diameter
1. Recognize the growth patterns of the normal trachea (rapid growth in diameter during the first two years after birth and at puberty)

b. Structure
1. Know the tracheal smooth muscle lies transversely between the dorsal tips of the tracheal rings
2. Understand the innervation of the tracheal smooth muscle
3. Understand the effects of contraction of the tracheal smooth muscle on airway diameter and stability
4. Understand the function of the mucus-secreting glands in the airway
5. Know that smooth muscle in the trachea is present as transverse bands between the dorsal tips of the cartilage rings

c. Cellular components (see cell biology)
1. Know how cilia beat in a metachronic way and propel mucus toward central airways
2. Know that the dynein arms contain ATPase necessary for ATP hydrolysis
3. Know the cell components of the airway epithelium (pseudostratified, columnar, ciliated epithelium, interspersed with goblet cells)
4. Recognize the normal structure of respiratory cilia on electron microscopy
5. Know the normal function of the cilia

6. Recognize the factors that affect mucociliary transport
7. Understand the role of mucociliary transport in airway clearance mechanisms

2. Bronchi
   a. Understand the difference in structure between bronchi and bronchioles
   b. Know that the total cross-sectional area of the airways increases exponentially with distance from the larynx
   c. Understand the actions of the subtypes of muscarinic receptors found within the airways
3. Bronchioles
   a. Know that there are no mucous glands in the bronchioles
   b. Understand the difference between terminal and respiratory bronchioles
4. Lungs
   a. Lobes
      1. Identify the lobes of the lung
      2. Know the distribution of the various bronchopulmonary segments
   b. Fissures
      1. Know which lobes constitute the boundaries of the major and minor fissures
5. Alveoli
   a. Understand the physiologic implications of the polyhedral shape of the alveoli
   b. Know that the alveolus has areas covered by a thin layer of fluid and that this "hypophase" is important in surfactant function
   c. Understand the factors that are responsible for alveolar stability
   d. Know that the epithelium and endothelium have a fused basement membrane on the thin side of the alveolar capillary membrane and that this facilitates gas transfer by reducing resistance to diffusion
   e. Know the components of the acinus (terminal respiratory units)
6. Collateral channels
   a. Know the definition of the pores of Kohn
   b. Know the definition of the canals of Lambert
   c. Understand the effects of age on the development of collateral channels (infants have poorly developed collateral channels)
7. Pulmonary lymphatics
   a. Understand the drainage pattern of the pulmonary lymphatics
   b. Know that the unidirectional flow of pulmonary lymph towards the hilus is maintained by valves in the lymphatic system
   c. Know that the function of the pulmonary lymphatics is to collect protein and water from the interstitium and return it to the circulation
   d. Know that the pulmonary lymphatics travel alongside the blood vessels
8. Innervation of the lung
   a. Know the innervation of airway smooth muscle
   b. Know the innervation of vascular smooth muscle
   c. Know the anatomy of sympathetic and parasympathetic innervation of the lung
   d. Understand the function of nonadrenergic/noncholinergic nervous system in the lung
   e. Know the putative mediators of the nonadrenergic/noncholinergic nervous system in the lung

f. Know that sensory nerves in the lung are vagal in origin

9. Blood vessels and circulation
    a. Pulmonary arteries
        1. Know that conventional pulmonary arteries run alongside the airways, and supernumerary pulmonary arteries do not run with the airways
        2. Know that there are two types of pulmonary arteries: conventional and supernumerary
        3. Understand the development of the muscularization of the pulmonary arterial system
        4. Understand the development of the response of the pulmonary arterial system to hypoxia (ie, increasing response with aging and extension of muscle)
        5. Know that pulmonary arteries can be classified as elastic, muscular, partially muscular, or nonmuscular
        6. Know that muscular arteries have a circular layer of muscle, bounded by internal and external elastic laminae
        7. Know that as arteries decrease in size, muscle becomes spiral and decreases in quantity
        8. Know that the endothelium of the pulmonary vascular system is continuous and nonfenestrated
        9. Know that endothelium is an active cell layer and not just a passive barrier
    b. Pulmonary veins
        1. Know that the pulmonary veins provide a reservoir for blood volume and help maintain a constant left ventricular output despite variable pulmonary blood flow
    c. Pulmonary circulation
        1. Recognize factors that regulate pulmonary circulation
        2. Recognize the effects of changing pleural pressure on pulmonary circulation
        3. Understand the contributions of extra-alveolar vessels and intra-alveolar vessels to pulmonary vascular resistance
    d. Systemic circulation
    e. Bronchial circulation
        1. Know that bronchial arteries extend into the lung to the level of the terminal bronchiole
        2. Know that there are numerous connections between the bronchial and pulmonary circulations
        3. Know that one third of the bronchial blood flow returns to the right atrium through the bronchial veins and the remainder to the left atrium via the pulmonary veins
        4. Know that bronchial arteries generally receive 1% to 2% of systemic blood flow

7. **Lung Defense Mechanisms**
    A. General
        1. Know the mechanisms that reduce the risk of developing pulmonary infections
    B. Modifications of inspired air
        1. Temperature

a. Recognize the conditions under which extrathoracic upper airways incompletely warm inspired air to body temperature

2. Humidification
   a. Know that there is a countercurrent mechanism for heating and humidification of inspired gas in the nose
   b. Know that inadequate humidification of inspired air impairs tracheal mucociliary transport
3. Particle deposition
   a. Know that particles smaller than 0.5 microns in diameter are not retained in the lung
   b. Know that particles 0.5 to 2 microns in diameter are preferentially deposited in the alveoli
   c. Know that particles 2 to 10 microns in diameter are preferentially deposited onto tracheobronchial epithelium
   d. Know that particles greater than 10 microns in diameter are preferentially deposited in the nasal passages during nasal breathing
   e. Know that nasopharyngeal filtering does not occur for particles less than 1 micron in diameter
   f. Know the factors that influence particle deposition in the lungs
   g. Know the role played by a spacer device in maximizing pulmonary deposition of an aerosol
4. Uptake of pollutant gases
   a. Know that the solubility of a gas will help determine the effects of the gas on the respiratory tract

C. Respiratory tract cilia
   1. Structure (see ciliary dysmotility, I.Q.2)
   2. Mechanism of action

D. Airway secretions
   1. Biochemical constituents
      a. Know that normal respiratory tract fluid is a mixture of secretions from submucosal gland ducts, goblet cells, and epithelial cells
      b. Know that the epithelial surface is covered by two layers: a low-viscosity periciliary fluid and a viscoelastic mucus layer
      c. Know that the viscosity and elasticity characteristics of normal tracheobronchial secretions are primarily due to the presence of mucous glycoproteins
      d. Know that the airway epithelium secretes antibacterial substances, including beta-defensins and lysozyme
      e. Know that surfactant proteins A and D have potent antimicrobial properties
      f. Know that airway epithelial cells regulate the depth of periciliary fluid
   2. Control of airway secretions
      a. Know that airway submucosal glands are under cholinergic nervous regulation (stimulation of cholinergic efferent nerves causes secretion from submucosal glands)
      b. Understand the effects of anticholinergic drugs upon submucosal gland secretion production in the airways

c. Know the histologic changes in the airways associated with chronic states of hypersecretion, eg, bronchitis, cystic fibrosis
d. Know the effect of beta-agonists on the secretion of periciliary fluid from airway epithelial cells

3. Mucociliary transport
a. Know that mucociliary mechanisms and alveolar macrophages both clear inhaled microorganisms
b. Understand the alterations in mucus that reduce mucociliary function
c. Know that mucociliary clearance is an important host defense in the nose, ears, and sinuses, as well as in the lower respiratory tract
d. Know the pharmacologic agents that alter mucociliary transport
e. Understand the factors that increase or decrease mucociliary transport

E. Cough
1. Normal physiology
a. Know that the highest densities of cough receptors are found in the larynx, trachea and bronchi
b. Know that chemical, mechanical, or pharmacologic stimulation of cough receptors causes afferent fibers in vagus nerve to transmit impulses to the medulla
c. Know the neural pathways important in cough
d. Understand the role of glottic closure in the production of effective cough
e. Understand the role of tracheal compression in the production of effective cough

2. Pathophysiology
a. Know the extrapulmonary afferent pathways that may produce cough
b. Recognize that muscular and efferent neural abnormalities may render cough ineffective

F. Respiratory reflexes and defense of the lung
1. Know the stimuli that can cause reflex laryngospasm and/or apnea in infants

G. Pulmonary lymphatics
1. Know that pulmonary lymphatics are an important component of the lung defense system
2. Know that efferent lymph vessels course through the connective tissue of the bronchovascular bundle and terminate in regional lymph nodes
3. Know the factors responsible for lung lymph fluid balance between secretion and absorption
4. Know the constituents of lung lymph fluid, including lymphocytes, protein, and lipids
5. Know the ontology of pulmonary lymph nodes

H. Air-blood barrier
1. Know that bronchoalveolar lavage cells constitute representative samples of cells present within alveoli and peripheral bronchioles
2. Know that tight junctions are a key defense in guarding alveoli against flooding

I. Pulmonary macrophage
1. Origin and distribution
a. Know that alveolar macrophages are derived from blood monocytes that replicate within the pulmonary interstitium and mature into macrophages

2. Function
a. Know that alveolar macrophages ingest and degrade foreign material

J. Cellular and humoral immunity
   1. Normal function
      a. Know that the secretory IgA in the upper airways is responsible for complement-independent neutralization of respiratory viruses
      b. Know that there are IgG subclasses
      c. Know that IgG is the major immunoglobulin within the alveoli
      d. Know that the major immunoglobulin within the upper airway is IgA
      e. Know the role of lysozyme in hydrolyzing structural components of bacterial cell walls
      f. Know that IgG subclass deficiency can predispose to infection, even when total IgG concentration is normal
      g. Know the role of neutrophils in the defense of the lung against bacterial, viral, mycobacterial, fungal, and parasitic diseases
      h. Know the role of lymphocytes in the defense of the lung against bacterial, viral, mycobacterial, fungal, and parasitic diseases
      i. Know the role of eosinophils in the defense of the lung against bacterial, viral, mycobacterial, fungal, and parasitic diseases
   2. Immunodeficiency states
      a. Understand the immunodeficiency states associated with pulmonary infection
      b. Know that recurrent infection may be the result of a defect in phagocytosis (chronic granulomatosis disease)
      c. Know that IgA deficiency is associated with atopy and airway reactivity
      d. Identify the immune deficiency state existing after lung trans- plantation and the organisms most likely to be pathogenic in that setting
      e. Know the manifestations of post-transplant lymphoproliferative disorder and that it is caused by Epstein-Barr virus (EBV)
      f. Know the roles of graft-versus-host disease and infection in the development of chronic lung disease after bone marrow transplantation

8. **Cell Biology and Biochemistry**

A. Cell and molecular biology
   1. Cell kinetics
      a. Know that the S (synthesis) stage is the period of DNA replication and the period of "labelling" with markers of proliferation (eg, tritiated thymidine, BrdU, PCNA)
   2. Cell differentiation
      a. Know the characteristics that distinguish progenitor cells from differentiated cells
   3. Protein synthesis
      a. Understand the relationships between DNA, mRNA, and amino acid sequences
      b. Recognize that protein expression may be modified at the levels of transcription, translation, and post-translation
      c. Understand the general principles of mRNA detection by Northern hybridization, microarray in situ hybridization, and reverse transcriptase polymerase chain reaction (RT PCR)
      d. Understand the general principles of protein detection by Western blot, immunoblot, 2-D gel electrophoresis, and immunohistochemical analyses
      e. Know that protein synthesis occurs in free ribosomes and in the rough endoplasmic reticulum

B. Lung cells and products
   1. Type I alveolar cell
      a. Know that type I and type II alveolar cells form tight junctions
      b. Know that type I alveolar epithelial cells cover more than 90% of alveolar surface although they are less numerous than type II epithelial cells
      c. Know that a type I cell arises from a type II cell and is a terminally differentiated cell incapable of division
   2. Type II alveolar epithelial cell
      a. Know the structural features of type II alveolar epithelial cells
      b. Know that type II alveolar cells secrete pulmonary surfactant
      c. Know that type II epithelial cells can divide
      d. Know that pulmonary alveolar epithelium reabsorbs edema fluid, in part by actively transporting ions
      e. Know the components of surfactant (ie, protein and phospholipids)
   3. Airway epithelial cell
      a. Understand the structural and functional differences between the apical and basolateral membranes of airway epithelial cells
      b. Know the function of tight junctions in airway epithelium
      c. Know that the principal site of nitric oxide production in the lower respiratory tract is the epithelial cell
   4. Endothelial cell
      a. Know that von Willebrand factor is synthesized by and is a characteristic feature of endothelium
      b. Understand the mechanisms by which the cell surface influences coagulation/fibrinolysis
      c. Know the actions of vasoactive compounds derived from endothelial cells
      d. Know the location and functions of angiotensin-converting enzyme
      e. Know which circulating compounds are metabolized by endothelium during their passage through the pulmonary circulation
   5. Alveolar macrophage
      a. Know that macrophages release cytokines
      b. Know the functions of alveolar macrophages
      c. Know the precursor cell of the alveolar macrophages
      d. Know the role of granulocyte-monocyte colony-stimulating factor (GM-CSF) and PU.1 on alveolar macrophage function
      e. Understand the role of alveolar macrophages in surfactant homeostasis
   6. Lymphocytes
      a. Identify lymphocyte subtypes and their functions
      b. Know that lymphocytes release cytokines
      c. Know the difference between a TH1 and a TH2 response
      d. Identify the major lymphocyte subsets by cell surface markers and their primary function (eg, CD4+ cells (T-helper cells))
   7. Leukocytes
      a. Know that the lung has large numbers of leukocytes and also is the predominant site of the "marginated pool" of leukocytes

b. Know that a degradation product of the fifth component of complement (C5) is involved in the generation of neutrophil chemotactic activity

8. Mast cell
   a. Know the functions of the mast cell
   b. Know the content of mast cell granules
   c. Understand the mechanism of mast cell activation
   d. Understand the mechanisms of granule and mediator release from mast cells
9. Mucous/goblet cell
   a. Know the stimuli that trigger release of products from mucous/goblet cells
10. Neuroendocrine (APUD) cells
   a. Know which vasoactive mediators and cytokines are released by neuroendocrine (APUD) cells
   b. Understand the relationship between the numbers of neuroendocrine (APUD) cells and neuroepithelial bodies and lung maturity or lung diseases characterized by chronic hypoxemia
11. Smooth muscle cells
   a. Understand mechanisms regulating smooth muscle tone
   b. Know that pulmonary vasculature smooth muscle in situ has a qualitatively different response to hypoxemia and acidosis relative to systemic vasculature
   c. Identify the mechanism of action and the effect of nitric oxide on vascular smooth muscle cells
12. Elastin
   a. Know that the elastin content of the lung increases with increasing gestational age
   b. Recognize the susceptibility of elastin to proteolysis by neutrophil elastase
   c. Know that the normal turnover rate of elastin is measured in years
13. Collagen
   a. Know different subtypes of collagen present in the normal and diseased lung (eg, fibrosing alveolitis)
   b. Recognize the susceptibility of collagen to proteolysis by neutrophil elastase
14. Proteinases/antiproteinases
   a. Identify major lung sources of proteinases and antiproteinases
   b. Know the activities of the major antiproteinases
   c. Know that reperfusion injury is caused by local generation of oxygen radicals
   d. Know that Clara cells have an important role in handling inhaled toxic substances
15. Oxidants and antioxidants
   a. Toxic oxygen radicals (see oxygen therapy)
      1. Know which toxic oxygen radicals are associated with hyperoxic lung injury
      2. Know that leukocytes and macrophages can generate toxic oxygen radicals
   b. Antioxidants in the lung
      1. Identify the major antioxidant enzymes and substrates within the lung, including catalase, superoxide dismutase, heme oxygenase, and glutathione peroxidase
16. Surface active materials
   a. Know the components of surface active materials
   b. Understand the functional role of surfactant-associated proteins
   c. Understand the functional role of surfactant lipids

d. Understand the developmental and hormonal regulation of surfactant production

17. Prostanoids
    a. Understand the actions of the arachidonic acid metabolites
    b. Know the pathways of arachidonic acid metabolism
    c. Know the mechanisms of action of leukotriene antagonist in asthma
18. Fibroblasts
    a. Know the anatomic location of fibroblasts in the lung
    b. Understand the role of fibroblasts in collagen synthesis and matrix formation
    c. Know the stimuli for fibroblast proliferation and collagen synthesis
19. Clara cells
    a. Know the anatomic location of Clara cells within the lung
    b. Know that two potential roles of Clara cells are surfactant recycling and active ion transport
20. Vitamin A
    a. Know that vitamin A deficiency can cause squamous metaplasia of the epithelium in large airways
21. Lysozyme
    a. Understand that lysozymes hydrolyze structural components of bacterial cell walls

9. **Gas Exchange, Ventilation-perfusion Distribution, Acid-base**

A. Gas exchange

1. Alveolar ventilation
    a. Effect on arterial PCO2
        1. Understand the relationship between minute ventilation and alveolar ventilation
        2. Understand the relationship between alveolar ventilation, carbon dioxide production, and arterial PCO2
    b. Effect on arterial PO2
        1. Calculate the effect of alveolar hypoventilation on alveolar and arterial PO2 using the simplified alveolar gas equation
2. Dead-space ventilation
    a. Identify the proportion of each tidal breath comprising anatomic dead space in conducting airways
    b. Calculate dead space/tidal volume ratios (Vd/VT) using the Bohr equation
    c. Recognize the limitations of end-tidal carbon dioxide as a measure of arterial PCO2
    d. Recognize the physiologic significance of an increased (PACO2-PaCO2) difference
    e. Understand how the physiologic dead space changes with body position (supine versus standing positions)
    f. Understand how exercise influences both minute ventilation and anatomic and physiologic dead-space ventilation
    g. Estimate the effect of the VD/VT ratio on alveolar ventilation
    h. Know the definition of respiratory dead space components (i.e., physiologic, anatomic, alveolar)
    i. Know that wasted ventilation does not affect arterial PO2 and PCO2
    j. Recognize technically adequate end-tidal carbon dioxide measurements as contrasted with poor end-tidal waveforms on a CO2 by time or volume plot

k. Know the pathophysiology of conditions leading to increases in alveolar dead

3. Ventilation-perfusion relationships in the lung
   a. Functional significance
      1. Know that V/Q mismatching is the most common reason for hypoxemia in patients with lung disease
      2. Understand how V/Q mismatching can lead to hypercapnia in addition to hypoxemia in the absence of compensatory hyperventilation
   b. Effect of gravity on gas distribution
      1. Recognize the factors that affect gas distribution within the lung
      2. Understand how change in body position (from supine to standing) alters ventilation-perfusion matching in healthy humans
      3. Understand how gravity affects the regional size of pulmonary structures and the regional minute ventilation within the lung
   c. Effect of gravity on pulmonary circulation
      1. Know the effect of gravity on the regional distribution of pulmonary flow
   d. Effect of gravity on ventilation/perfusion
      1. Understand the effect of gravity on regional ventilation/perfusion ratio
      2. Understand the effect of gravity on regional PA02 and PAC02
4. Oxygen consumption and carbon dioxide production
   a. Measuring oxygen consumption and carbon dioxide production
      1. Know how to measure and calculate oxygen consumption by expired gas analysis during rest or exercise
      2. Know how to measure and calculate CO2 production by expired gas analysis during rest or exercise
   b. Respiratory exchange ratio
      1. Recognize the clinical features that increase oxygen consumption and carbon dioxide production in children
      2. Understand that acute hyperventilation (anxiety) can increase the respiratory exchange ratio
   c. Influence of carbohydrate, fat, and protein
      1. Know the respiratory quotient of fat, carbohydrate, and protein
      2. Know that carbohydrate ingestion is associated with an increased respiratory quotient secondary to increased CO2 production
5. Alveolar air equation
   a. (PAO2-PaO2) difference
      1. Calculate how a difference in barometric pressure affects arterial PO2
      2. Calculate each of the factors in the alveolar air equation
      3. Know that normal values for the (PAO2-PaO2) difference change with the fraction of inspired oxygen in healthy individuals
      4. Know the definition of alveolar ventilation
      5. Know the conditions associated with an increased a-A PO2
      6. Know the approximate reduction in alveolar partial pressure of oxygen with increases in altitude in the absence of hyperventilation (i.e., 5 mm Hg per 1000 feet ascent)
      7. Know that even pressurized commercial aircraft have cabin pressures compatible with altitudes of approximately 8000 feet and be able to estimate the

reduction in alveolar oxygen (40 mm Hg) that could occur from sea level to stable flight

b. PaO2:PAO2 ratio
    1. Know that the PaO2/PAO2 ratio does not change with the fraction of inspired oxygen in healthy humans
    2. Know the normal value for the PaO2/PAO2 ratio and how this differs in newborn infants
    3. Understand the relationship between PCO2 and pH in arterial blood
    4. Know the relationship between PaO2 and O2 content

6. Fick's law of diffusion
    a. Know the determinants of gas diffusion through a membrane according to Fick's first law of diffusion
    b. Know why carbon monoxide is used as the inspiratory gas to measure diffusing capacity of the lung
7. Arterial hypoxemia
    a. Understand the effect of intrapulmonary shunt on arterial PO2
    b. Know that hypercapnia does not change the (PAO2-PaO2) difference for oxygen
    c. Recognize that pure or uncomplicated upper airway obstruction is associated with a normal (PAO2-PaO2) difference
    d. Understand how mixed venous oxygen content contributes to arterial hypoxemia in lungs with V/Q mismatching
    e. Recognize the pulmonary conditions that produce right-to-left shunts in children with hypoxemia
    f. Know that hypoxemia due to gas diffusion limitation is most likely to occur during exercise in patients with lung disease or at high altitudes
    g. Distinguish ventilation-perfusion imbalance from anatomic shunt on the basis of blood gas data
    h. Know the four physiologic causes of hypoxemia
    i. Know how hypoventilation causes hypoxemia
8. Shunt equation
    a. Calculate shunt fraction from the shunt equation
    b. Know the anatomic cause of shunt in healthy individuals
9. Oxygenation/oxygen-carrying capacity
    a. Calculate oxygen-carrying capacity and understand its clinical application
    b. Recognize the clinical factors that alter oxygen-carrying capacity
    c. Know the adaptations to deficiencies in oxygen-carrying capacity
    d. Know the definition of venous admixture
10. Oxygen transport
    a. Diffusion barriers
        1. Know that increasing FIO2 in normal lungs does not increase oxygen diffusion into arterial blood
    b. Oxyhemoglobin dissociation curve
        1. Recognize the factors that change the shape of the oxyhemoglobin dissociation curve
        2. Recognize the factors that shift the oxyhemoglobin dissociation curve

3. Recognize the changes in arterial PO2 and P50 associated with methemoglobinemia
4. Recognize that some hemoglobinopathies (eg, sickle cell disease) affect the shape and/or position of the oxy-hemoglobin dissociation curve

c. Capillary transit time of erythrocytes
1. Recognize the factors affecting the pulmonary capillary transit time of erythrocytes

d. Oxygen delivery to tissues
1. Recognize the factors that influence oxygen transport to the tissues
2. Understand the relationship between oxygen delivery and oxygen consumption in healthy individuals

11. Oxygen exchange in tissues
a. Recognize local factors at the tissue level that ensure adequate oxygen supply to a given cell

B. Carbon dioxide transport and acid-base balance
1. Factors affecting transport
a. Recognize the factors affecting carbon dioxide transport
b. Know how carbon dioxide is carried in blood

2. Hydration of carbon dioxide
a. Carbonic anhydrase
1. Know that carbonic anhydrase is located in vascular endothelium and in erythrocytes
2. Know that carbonic anhydrase accelerates the hydration of carbon dioxide
3. Know that carbonic anhydrase inhibitors can be used to treat altitude sickness by stimulating ventilation

b. Henderson-Hasselbalch equation
1. Understand the Henderson-Hasselbalch equation

c. Buffering of hydrogen ion
1. Know the major intracellular and extracellular hydrogen ion buffers
2. Know the rationale for correcting the base excess value

d. Chloride shift in erythrocytes

C. Excretion of acid
1. Normal
a. Lungs
1. Recognize factors limiting excretion of acid by the lungs

b. Kidneys
1. Recognize factors that influence excretion of acid by the kidneys

2. Respiratory acidosis (acute and chronic)
a. Causes
1. Recognize the blood gas and pH changes that are caused by voluntary breath holding
2. Recognize acute respiratory acidosis vs chronic respiratory acidosis by arterial blood gas analysis
3. Identify metabolic compensation of respiratory acidosis and understand that this implies chronic respiratory acidosis

b. Treatment

c. Diagnosis
    1. Recognize the implications of the Davenport diagram and blood buffer line in the interpretation of arterial blood gas values
3. Respiratory alkalosis (acute and chronic)
    a. Causes
        1. Understand that respiratory alkalosis can be caused by acute hyperventilation
    b. Treatment
        1. Know the therapy for acute respiratory alkalosis (hyperventilation)
    c. Diagnosis
        1. Recognize manifestations of salicylate intoxication on arterial blood gas analysis
4. Metabolic acidosis (acute and chronic)
    a. Causes
        1. Recognize the clinical causes of metabolic acidosis (renal, cardiovascular, gastrointestinal, ingestion, intravenous fluid therapy)
        2. Be able to distinguish between an anion gap metabolic acidosis and a nonanion gap (hyperchloremic) metabolic acidosis
        3. Be able to describe the causes and basic pathophysiology of both iatrogenic and endogenous hyperchloremic metabolic acidosis
        4. Be able to describe the causes and simple pathophysiology of anion gap metabolic acidosis (eg, methanol, uremia, diabetic ketoacidosis, paraldehyde, iron, isoniazid (INH), lactic acid, ethanol, ethylene glycol, salicylates)
    b. Treatment
        1. Recognize the effects of sodium bicarbonate administration on CO2 production in patients with lung disease
        2. Recognize the risks of bicarbonate administration in metabolic acidosis
    c. Diagnosis
5. Metabolic alkalosis (acute and chronic)
    a. Causes
        1. Recognize the role of chloride loss in causes of metabolic alkalosis (eg, cystic fibrosis, diuretics)
        2. Know the causes of metabolic alkalosis, including chronic diuretic therapy and chronic hypercapnia
        3. Understand the role of the renin-angiotensin-aldosterone system in maintaining a contraction alkalosis
    b. Treatment
        1. Know how to treat metabolic alkalosis
        2. Know when to treat metabolic alkalosis (i.e., patients on digitalis preparations, with low ionized calcium and/or magnesium, with low potassium, with ventilatory drive suppression)
    c. Diagnosis
        1. Recognize manifestations of metabolic alkalosis on arterial blood gas analysis
6. In vivo vs in vitro CO2 dissociation curve

D. Exercise physiology (see II.C.)

10. **Respiratory Mechanics**

A. Static respiratory system mechanics

1. Definitions
    a. Know the definitions of and the factors that determine the various lung volumes and capacities
    b. Know the difference between lung compliance and total respiratory system compliance
    c. Know the definition of compliance
    d. Recognize the difference between static and dynamic compliance
    e. Know the definition of frequency dependence of compliance
    f. Know that specific compliance or elastance is the measured value normalized by another measurement, usually lung volume
    g. Know that compliance is the slope of a pressure- volume curve and varies as a function of lung volume
    h. Recognize that dynamic compliance decreases with increasing respiratory frequency in subjects with airway obstruction
    i. Know the definition of chest wall compliance
    j. Know how compliance varies as a function of lung volume
    k. Know the changes in specific compliance through childhood in normal children
2. Surface tension (see VIII.O)
    a. Know the definition of surface tension
    b. Know the method of measuring surface tension
    c. Understand the LaPlace relationship as it applies to alveolar curvature, alveolar surface tension, and lung recoil
    d. Know that pulmonary surfactant lowers surface tension to a greater extent when the alveolar surface is being compressed than when it is expanded (hysteresis)
    e. Understand how surfactant stabilizes alveoli of different sizes by changing the surface tension at the alveolar air-liquid interface
3. Elastic recoil of the lung
    a. Recognize the pressure-volume curve of the normal lung
    b. Know that the pressure-volume characteristics of the normal lung reflect surface forces and tissue recoil
    c. Know that the inflation and deflation pressure-volume curves of the lung differ (hysteresis)
    d. Know that the normal lung is relatively stiff at total lung capacity and relatively compliant at functional residual capacity
    e. Recognize factors that influence lung compliance in health and disease
    f. Know how to measure static and dynamic pulmonary compliance
    g. Know the relative contributions of surface forces and tissue forces to lung recoil
4. Static mechanics of the chest wall
    a. Know the pressure-volume characteristics of the chest wall of a normal child
    b. Know how the pressure-volume curve of the chest wall of an infant differs from that of the older child or adult
    c. Know that a normal pressure-volume curve of the chest wall assumes that the respiratory muscles are relaxed
    d. Know that the chest wall is relatively stiff at low lung volumes and relatively compliant at volumes above functional residual capacity
    e. Recognize factors that influence chest wall compliance in health and disease

f. Know the functional sequelae that may be associated with a highly compliant chest wall

5. Static mechanics of the combined lung & chest wall
   a. Know how the pressure-volume curve of the lung and that of the chest wall are combined to describe the mechanical characteristics of the respiratory system
   b. Know that end-expiratory volume in infants is actively determined
   c. Be able to calculate total respiratory system compliance
6. Pleural pressure
   a. Understand the relationships among pleural pressure, transpulmonary pressure, static alveolar pressure, and the pressure drop across the chest wall
   b. Know that intrapleural pressure in a patient who is at rest and breathing quietly is negative compared to atmospheric pressure throughout the breathing cycle
   c. Know how intrapleural pressure varies topographically within the thorax in a gravitational field
   d. Know that esophageal pressure approximates pleural pressure
   e. Know methods of measuring esophageal pressure
   f. Know that chest wall distortion, esophageal muscle contraction, and cardiac motion influence esophageal pressure independent of changes in pleural pressure

B. Airway mechanics
1. Definitions
   a. Know the definitions of resistance and conductance
   b. Know the definitions and determinants of frequency dependence of resistance
   c. Understand how turbulent and laminar flow regimes influence airway resistance
   d. Understand how gas density and viscosity influence airway resistance during turbulent and laminar flow
   e. Understand the relationship between resistance and radius in a rigid tube under laminar flow conditions
   f. Know how resistance and compliance of a system determine the rate at which the system will empty passively (the time constant)
   g. Understand the functional significance of the expiratory time constant
   h. Know that increased flow rates can require a geometric increase in driving pressure under turbulent conditions (i.e., that resistance is not constant)
   i. Know that a helium-oxygen gas mixture can be used to reduce the work of breathing in airway obstruction where flow is highly turbulent (e.g., croup, tracheal narrowing)
   j. Know that a time constant of a lung region can be calculated as its resistance times it compliance
   k. Understand that lung regions with the shortest time constants fill first on inspiration and empty first on expiration and that they are generally the best ventilated lung regions
   l. Understand how pulmonary time constants impact optimal mechanical ventilator strategies in patients with obstructive or restrictive lung disease
2. Airway resistance/conductance
   a. Understand the relative contributions of the upper airway, central tracheobronchial tree, and peripheral airways to total airway resistance

b. Understand the difference between airway resistance and total respiratory system resistance
c. Know various methods of measuring airway and respiratory system resistance
d. Understand how airway resistance varies as a function of lung volume
e. Know changes in specific airway conductance during the first year after birth

3. Maximal flows/flow limitation
   a. Know that maximal expiratory flow over much of the vital capacity range is relatively effort independent
   b. Appreciate the relationship between the maximal expiratory flow-volume curve and the forced spirogram
   c. Know that maximal expiratory flow at a given lung volume is a function of airway geometry, airway wall compliance, and lung recoil
   d. Appreciate that maximal expiratory flow is a function of lung volume over most of the vital capacity range, while maximal inspiratory flow is relatively independent of lung volume
   e. Appreciate that with increasing expiratory flow, pressure within the airways becomes lower than pleural pressure and the airways therefore narrow
   f. Understand how maximal expiratory flow varies as a function of gas density and viscosity
   g. Understand the basic mechanisms of maximum expiratory flow limitation
   h. Know that obstructive diseases causing a heterogeneous distribution of time constants in the lung will result in a flow volume plot that is convex to the volume axis (i.e., 'scooped out' in appearance)

C. Work of breathing
   1. Know that the work of breathing is related to the area of a dynamic pressure-volume curve
   2. Know the components (elastic and resistive) of the work of breathing
   3. Know how the optimal respiratory rate/tidal volume combination for a given minute ventilation changes in obstructive lung disease
   4. Know how the optimal respiratory rate/tidal volume combination for a given minute ventilation changes in restrictive lung disease
   5. Know the equation of motion of the lung over normal breathing frequencies

11. **Control of Breathing**

A. Peripheral receptors
   1. Chemoreceptors
      a. Carotid bodies
         1. Location
            a. Know the location of the carotid bodies
         2. Innervation
            a. Know the primary afferent innervation of the carotid bodies
         3. Structure
            a. Know that the carotid body has a very high blood flow per gram of tissue
            b. Know the blood supply of the carotid bodies
         4. Physiology
            a. Know that the peripheral chemoreceptors are the main arterial oxygen sensors in mammals

b. Know that peripheral chemoreceptors exhibit tonic activity, even at physiologic arterial PO2
c. Know that peripheral chemoreceptor activity increases exponentially at arterial PO2 less than 60 mm Hg
d. Know that carotid bodies respond to large changes in arterial pH
e. Understand the factors that influence the peripheral chemoreceptor response to PO2 (i.e., increased arterial PCO2)
f. Know that the peripheral chemoreceptor responds to arterial PO2 and not to oxygen content
g. Know that the peripheral chemoreceptor responds to large increases in PCO2
h. Know that hypercapnia and acidosis amplify the peripheral chemoreceptor-stimulated ventilatory response to hypoxia synergistically, while hypocapnia and alkalosis depress it

b. Aortic bodies
1. Know that the aortic bodies play a role in the redistribution of the fetal circulation during fetal hypoxemia
2. Know that the aortic bodies respond mainly to arterial oxygen content and not to arterial PO2

c. Laryngeal chemoreceptors
1. Location
a. Know that laryngeal chemoreceptors are located primarily on the laryngeal surface of the epiglottis
2. Innervation
a. Know the main afferent innervation of the laryngeal chemoreceptors (i.e., superior laryngeal nerves)
3. Stimuli
a. Recognize the stimuli that can activate laryngeal chemoreceptors
4. Reflex responses
a. Recognize the reflex responses produced by stimulation of laryngeal chemoreceptors and understand their effects on regulation of breathing

2. Mechanoreceptors
a. Vagal
1. Pulmonary stretch receptors
a. Location
1. Explain the role of mechanoreceptors in the control of ventilation
2. Know the location of slowly adapting stretch receptors
3. Know the location of rapidly adapting irritant receptors
4. Know the innervation of the stretch receptors (militated vagal afferent nerves)
5. Know the innervation of irritant receptors (militated vagal afferent nerves)
b. Stimulus
1. Know that increasing lung volume is the stimulus for slow- adapting stretch receptors

2. Know that both mechanical and chemical stimuli elicit responses from the rapidly adapting irritant receptors

c. Reflex response

1. Know that the Hering-Breuer reflex is mediated by slowly adapting stretch receptors
2. Know that cough, bronchoconstriction, and increased mucus production are responses to stimulation of rapidly adapting irritant receptors
3. Know that hyperpnea is the dominant response to stimulation of rapidly adapting stretch receptors within the lung
4. Understand the role of irritant receptors in the adaptive response to adverse environmental stimuli

2. J receptors

a. Know the anatomic location of the J receptors
b. Know the stimuli for J-receptor activity
c. Recognize the pattern of ventilatory response to stimulation of the J receptors

b. Chest wall receptors

1. Types

a. Know that muscle spindles innervated by gamma-afferent fibers are found in the intercostal muscles

2. Role

a. Know that muscle spindles in the intercostal muscles are involved in respiratory compensation for increased work of breathing

B. Central chemoreceptor

1. Location

a. Know that central chemoreceptors are located in the medulla oblongata
b. Know that central chemoreceptors are separated from arterial blood

2. Stimulus

a. Recognize the pattern of response of the central chemoreceptors to increased arterial $PCO_2$
b. Know that the central chemoreceptor accounts for more of the ventilatory response to carbon dioxide than the carotid bodies
c. Know that central chemoreceptors respond to pH changes in cerebrospinal fluid (CSF) surrounding the medulla: arterial $CO_2$ diffuses into the CSF, stimulating the chemoreceptors, which increases ventilation
d. Know that central chemoreceptors stimulate a linear increase in minute ventilation as $PaCO_2$ is increased
e. Know that central chemoreceptor stimulation can be blunted by chronically increased $PaCO_2$ as metabolic compensation restores blood pH towards normal
f. Know that hypoxemia and acidemia accentuate the central chemoreceptor-stimulated ventilatory response to $CO_2$ while hyperoxemia and alkalemia depress it

C. Central neuronal control

D. Factors that influence control of respiration

1. Development of control of respiration

a. Fetal breathing activity

1. Identify factors that increase and decrease fetal breathing

2. Know that fetal breathing activity occurs only during REM sleep and is present about 40% of the time

b. Hypoxic drive
1. Know that there is a maturation of hypoxic drive during infancy

c. Hypercapnic drive
1. Understand factors that influence hypercapnic ventilatory response
2. Understand the interaction of the hypoxic and hypercapnic drives

d. Periodic breathing
1. Know that hypoxemia can lead to periodic breathing in a healthy infant
2. Know that periodic breathing may occur in normal infants
3. Realize when periodic breathing is abnormal in infants

e. Central Apnea
1. Know that short central apnea is normal in children of all ages
2. Know that prolonged apnea and bradycardia in preterm infants typically resolve by about 43 weeks post-conceptional age

2. Sleep

a. NREM sleep
1. Pattern of breathing
a. Know that NREM sleep is generally characterized by regular breathing
2. Control of breathing
a. Know that chemical/metabolic stimuli control ventilatory rate and depth during NREM sleep
b. Know that the slope of the ventilatory response curve is decreased during NREM sleep compared with the response during the awake state

b. REM sleep
1. Pattern of breathing
a. Know that paradoxical inward rib cage motion during inspiration is a normal characteristic of breathing during REM sleep in infants
b. Know that an irregular breathing pattern is characteristic of REM sleep
2. Control of breathing
a. Be able to compare the effects of REM sleep and NREM sleep on the ventilatory response to carbon dioxide
b. Know that in REM sleep, as compared with NREM sleep, chemical control is less important and behavioral (cortical) control is of increased importance
3. Effect on postural muscles
a. Recognize the effect of REM sleep on muscle tone of the postural muscles and upper airway muscles

E. Measurement of respiratory drive

1. Techniques
a. Understand the technique for ventilatory response testing in children

2. Normal response
a. Recognize the pattern of the normal response to hypercapnia
b. Recognize the factors that modify the normal response to acute hypoxia
c. Recognize the effects of chronic high altitude exposures on respiratory drive
d. Know the acute ventilatory responses to high altitude

e. Recognize that the normal ventilatory response to hypoxia is an exponential function of PaCO2 and a linear function of SaO2

3. Abnormal response

12. **Pulmonary Vascular Physiology**

A. Pulmonary vascular physiology

1. Recognize the effect of lung volume on pulmonary vascular resistance
2. Know the physiologic determinants of Zone I, II, and III conditions in the lung
3. Describe the physiologic changes in pulmonary circulation that occur at birth
4. Recognize the factors that cause vasodilation and vasoconstriction of the pulmonary vasculature
5. Describe the normal transvascular fluid flow within the lung
6. Understand the effects of body position (ie, standing versus recumbent) on distribution of pulmonary blood flow
7. Understand how hypoxic pulmonary vasoconstriction is modified by pH
8. Know that hypoxic pulmonary vasoconstriction is not linearly related to alveolar PO2, with increased constriction developing with a PaO2 below 60 mm Hg
9. Recognize the factors that cause pulmonary vasoconstriction
10. Recognize the factors that dictate pulmonary artery pressure
11. Recognize the changes that occur in the pulmonary circulation with exercise
12. Understand where nitric oxide is produced and how it affects pulmonary vascular tone
13. Understand why inhaled nitric oxide may be effective in improving oxygenation and lowering pulmonary vascular resistance in patients with acute respiratory distress syndrome
14. Know that the pulmonary vascular system is a low-resistance, low-pressure, high-compliance vascular bed
15. Describe the influence of alveolar surface tension on transvascular fluid balance
16. Recognize the changes that occur in the pulmonary circulation in an atelectatic lobe and their impact on the time course of related arterial hypoxemia
17. Describe the factors affecting transvascular fluid flow within the lung

B. Bronchial vascular physiology

1. Physiology of bronchial circulation
   a. Understand the factors that influence blood flow through the bronchial vasculature
   b. Understand the effect of vasoactive compounds on the bronchial vasculature
   c. Understand how chronic lung inflammation due to bronchiectasis may lead to bronchial artery hypertrophy with subsequent hemoptysis

13. **Mechanisms of Lung Inflammation, Injury and Repair**

A. Pulmonary inflammation

1. Components of an inflammatory response
   a. Vasodilation
      1. Know that vasodilation is one of the components of an acute inflammatory reaction
   b. Altered permeability
      1. Know the relative permeability of endothelium (as compared with epithelium) in the alveoli during acute inflammatory processes
   c. Leukocytic infiltration
      1. Acute

a. Know the types of cell that accumulate in an acute inflammatory reaction
b. Recognize an acute inflammatory response within the lung (on histology)

2. Chronic
a. Know the time course of accumulation of mononuclear phagocytes in an inflammatory reaction
b. Recognize a granulomatous inflammatory response within the lung (on histology)

2. Initiation of inflammation
a. Stimuli of inflammation
1. Immunologic
a. Understand the role of cytotoxic T-cells in the development of graft rejection and viral pneumonitis
b. Understand the relative contributions of specific antibodies, antigen-antibody complexes, and sensitized T-cells on the pathophysiology of common immunologic lung diseases
c. Understand the mechanism of action of immune suppressive drugs used in patients who have had a lung transplant

2. Nonimmunologic
a. Identify the products of airway epithelial cells that can contribute to airway inflammation
b. Recognize the factors that contribute to neutrophil chemotaxis in the lung
c. Know the mechanisms responsible for squamous metaplasia of the airway epithelium

b. Chemical mediators of inflammation
1. Vasodilation
a. Know which mediators of inflammation are vasodilators

2. Altered permeability

3. Cellular infiltration
a. Know that proteolytic degradation of C3 and C5 generate neutrophil chemotactic factors
b. Know that an early feature of airway damage is impairment of ciliary function
c. Know that IL-8 and LT-B4 are potent neutrophil chemoattractant factors
d. Identify the role of IL-5 as an eosinophil chemoattractant

4. Mediators that control inflammation
a. Know which mediators reduce the inflammatory response (eg, soluble TNF receptors, IL-10, etc)
b. Know that exhaled nitric oxide is increased in the presence of asthmatic inflammation
c. Identify the roles of IL-4, IL-13, and LT-D4 in asthmatic inflammation

3. Cells - actions/interactions
a. Granulocytes
1. Neutrophils and their products
a. Know the mechanism by which neutrophil elastase causes lung injury

2. Eosinophils and their actions
a. Know the mechanisms of eosinophil-mediated lung inflammation

b. Mononuclear phagocytes
    1. Understand the role of mononuclear phagocytes in terms of antigen presentation to lymphocytes
    2. Identify the inflammatory products of macrophages
    3. Recognize the actions of the more common cytokines
c. Lymphocytes
    1. Recognize the actions of the more common lymphokines
    2. Know the role of type 1 T-helper cells in nonallergic inflammatory reactions
    3. Know the role of type 2 T-helper cells in allergic inflammatory reactions

B. Mechanisms of injury
1. Oxygen radicals (see VIII, Oxygen therapy)
2. Proteases (see VIII and alpha1-antitrypsin)
3. Other (see VIII and I.N. ARDS)

C. Repair
1. Normal
    a. Understand the role of type II alveolar cells in normal repair of inflammation
2. Abnormal
    a. Recognize factors that may lead to fibrosis within the lung
    b. Know that macrophages and activated epithelial cells promote fibrosis

14. **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