SECTION 8

PULMONARY MEDICINE
CHAPTER 66

EVALUATION OF CHRONIC COUGH

1. EPIDEMIOLOGY
   - Nearly all adult cases of chronic cough in nonsmokers who are not taking an ACEI can be attributed to the “Pathologic Triad of Chronic Cough” (asthma, GERD, upper airway cough syndrome [UACS; previously known as postnasal drip syndrome]).
   - ACEI cough is idiosyncratic, occurrence is higher in female than males

2. PATHOPHYSIOLOGY
   - Afferent (sensory) limb: chemical or mechanical stimulation of receptors on pharynx, larynx, airways, external auditory meatus, esophagus stimulates vagus and superior laryngeal nerves
   - Receptors upregulated in chronic cough
   - CNS: cough center in nucleus tractus solitarius
   - Efferent (motor) limb: expiratory and bronchial muscle contraction against adducted vocal cords increases positive intrathoracic pressure

3. DEFINITION
   - Subacute cough lasts between 3 and 8 weeks
   - Chronic cough duration is at least 8 weeks

4. DIFFERENTIAL DIAGNOSIS
   - Respiratory tract infection (viral or bacterial)
   - Asthma
   - Upper airway cough syndrome (postnasal drip syndrome)
   - CHF
   - Pertussis
   - COPD
   - GERD
   - Bronchiectasis
   - Eosinophilic bronchitis
   - Pulmonary tuberculosis
   - Interstitial lung disease
   - Bronchogenic carcinoma
   - Medication-induced cough

5. EVALUATION AND TREATMENT OF THE COMMON CAUSES OF CHRONIC COUGH
   - Upper airway cough syndrome: rhinitis, sinusitis, or postnasal drip syndrome
   - Presentation: symptoms of rhinitis, frequent throat clearing, itchy throat or palate, although some are asymptomatic; exam may reveal edematous nasal turbinates and a glistening “cobblestone” appearance of the oropharynx
     - Empiric trial of an oral antihistamine
     - Nasal corticosteroids × 3–4 weeks
     - Ipratropium nasal spray can be added for refractory cough
     - Consider empiric antibiotics if coronal CT scan of sinuses suggests sinusitis
Evaluation of Chronic Cough

- Cough-variant asthma: atopic history or family history of eczema, allergies, or asthma; history of cough triggers (e.g., exercise, cold exposure, environmental allergens, or animal dander); only manifestation of asthma in up to 55% of pts
  - Test with routine spirometry, and, if normal, a methacholine challenge test; if both are normal, they effectively rule out cough-variant asthma
  - Consider an empiric trial of inhaled steroids and albuterol for 6–8 weeks
  - Leukotriene receptor antagonists can be added for refractory symptoms
- Gastroesophageal reflux (GERD): history of heartburn, dyspepsia, or sour taste in the mouth exacerbated by meals and supine position; also, may have frequent throat clearing, morning hoarseness, and a globus sensation
  - Up to 75% of patients with GERD-induced cough have no reflux symptoms
    - Nonpharmacologic interventions: diet high in protein; avoid bedtime snacks, fatty foods, chocolate, excess alcohol, caffeine, mints, and citrus fruits; smoking cessation and elevate head of the bed 6 inches
    - Empiric trial of proton pump inhibitor or moderate–high dose H₂-blockers for 4–6 months; extend therapy for 3 months past resolution of symptoms
    - Metoclopramide 10 mg PO after meals and at bedtime can be added for refractory cough
    - Insufficient data to support Nissen fundoplication for refractory cases
    - Esophageal pH probe testing usually not necessary but solidifies diagnosis

6. EVALUATION OF LESS COMMON CAUSES OF CHRONIC COUGH

- Initial studies and interventions to consider
  - Chest X-ray, place a PPD test, and stop ACEI therapy
  - Investigate for toxic occupational exposures and counsel to stop smoking
  - Pulmonary function tests to assess for chronic bronchitis or cough asthma
  - Coronal CT scan of paranasal sinuses to rule out chronic sinusitis
  - Induced sputum for eosinophils greater than 3% and normal methacholine challenge test indicate eosinophilic bronchitis; treat with inhaled steroids × 14 days
- Second-tier diagnostic studies
  - High-resolution CT scan of chest to evaluate for interstitial lung disease or bronchiectasis if chest X-ray abnormal and high clinical suspicion
  - Bronchoscopy indicated if high suspicion for lung cancer or foreign body
- See Figure 66.1
FIGURE 66.1. Evaluation of Chronic Cough in Immunocompetent Patients

Source: Adapted from Chest, 1998; 114 (Suppl 2 Managing): 166S.

REFERENCES

CHAPTER 67
EVALUATION OF DYSPNEA

1. PATHOPHYSIOLOGY
   • Afferent (sensory) limb: feedback from peripheral receptors to sensory cortex
     ▪ Decreased PaO₂, increased PaCO₂, decreased pH all stimulate chemoreceptors in carotid bodies and medulla
     ▪ Bronchospasm stimulates mechanoreceptors in lung
     ▪ Interstitial fluid, increased PA pressure triggers pressure receptors in pulmonary vasculature
     ▪ Exercise stimulates metaboreceptors in skeletal muscle
   • Efferent (motor) limb: feed-forward from motor cortex to ventilatory muscles
   • CNS: respiratory center in medulla
   • Mismatch between feedback and feed-forward signals increase dyspnea

2. ETIOLOGIES
   • Common etiologies
     ▪ Asthma: intermittent breathlessness, certain triggers, allergic rhinitis, prolonged expiration, wheezing
     ▪ COPD: history of smoking, barrel chest, prolonged expiration, wheezing
     ▪ CHF: history of HTN, CAD, or DM; orthopnea, paroxysmal nocturnal dyspnea, pedal edema, JVD, bibasilar rales, wheezing, S3 gallop
     ▪ Anxiety: history of anxiety, PTSD, OCD, panic disorder; sighing breathing
     ▪ GERD: postprandial dyspnea
   ▪ Hemoptysis suggests cancer, pneumonia, bronchiectasis, arteriovenous malformation
   ▪ Recurrent pneumonia suggests lung cancer, bronchiectasis, aspiration
   ▪ Drug exposure: β blockers can exacerbate reactive airway disease; amiodarone and nitrofurantoin can cause pneumonitis; methotrexate can cause lung fibrosis
   ▪ Immunosuppression: consider opportunistic infections including PCP, tuberculosis, legionella, cytomegalovirus, aspergillus, and coccidiomycosis

3. DIFFERENTIAL DIAGNOSIS
   • Panic attack
   • Pneumonia
   • COPD
   • Interstitial lung disease
   • Asthma
   • Pneumothorax
   • Pulmonary embolus
   • CHF
   • Acute myocardial infarction
   • Arrhythmia
   • Metabolic acidosis
   • Cyanide toxicity
   • Methemoglobinemia
   • Carbon monoxide poisoning
   • Conversion disorders
   • Malingering
4. WORKUP
- Initial testing
  - CBC, chemistry panel, chest radiograph, ECG, spirometry, pulse oximetry
  - Treat common causes (asthma, COPD, CHF, pleural effusion, anemia) accordingly
- Secondary testing
  - Echocardiogram, BNP, pulmonary function testing, arterial blood gas, Holter monitor, ventilation-perfusion scan, high-resolution CT scan, myocardial perfusion cardiac study
  - Treat cause accordingly (pericardial disease, CHF, valvular heart disease, CAD, arrhythmia, restrictive lung disease, interstitial lung disease, chronic PE)
- Tertiary testing
  - Cardiac catheterization, cardiopulmonary exercise testing, bronchoscopy, esophageal pH testing, open lung biopsy
  - Treat cause accordingly (GERD, CAD, deconditioning, pulmonary hypertension, psychogenic dyspnea)

5. TREATMENT
- Treat accordingly based on etiology of dyspnea

REFERENCE
CHAPTER 68
EVALUATION OF HEMOPTYSIS

1. EPIDEMIOLOGY
   • United States: bronchitis 50%, primary lung cancer 23%, bronchogenic carcinoma 5–44% (20% develop hemoptysis at some point, but only 7% at initial diagnosis), idiopathic 7–34%
   • Worldwide: Tb is the most common cause with prevalence of 2 billion people
   • Massive hemoptysis, whether in developed or developing countries, is attributed to Tb, mycetoma, or lung abscess

2. PATHOPHYSIOLOGY
   • Anatomic approach
   • Alveolar (diffuse alveolar hemorrhage): capillaries overloaded by pulmonary circulation (low pressure)
   • Inflammatory: small-vessel vasculitis (granulomatosis with polyangitis [formerly known as Wegener’s granulomatosis], microscopic polyangiitis, SLE, Goodpasture’s syndrome, post-BMT)
   • Noninflammatory: inhalational injury (burns, cocaine, toxins)
   • Small and medium airways: bronchial vessels from systemic circulation (high pressure)
   • Infectious: bronchitis, bronchiectasis, pneumonia (especially cavitary), tuberculosis, lung abscess, paragonimiasis
   • Noninfectious: inhalation, trauma, foreign body, lung cancer (especially in proximal airways, which leads to erosion into hilar vessels), metastases
   • Pulmonary vessels: increased LA pressure from CHF, MR with focal regurgitant jet, AVMs, PE
   • Pulmonary hypertension

3. CLINICAL PRESENTATION
   • Hemoptysis: spitting of blood from lungs or bronchi
   • Historical clues
     • Fever, productive cough suggests upper respiratory infection, pneumonia
     • Dyspnea on exertion with orthopnea suggests CHF
     • Pleuritic chest pain suggests pulmonary embolus
     • Anticoagulant use suggests medication effect
     • History of breast, colon, or renal cancer suggests metastatic disease
     • History of chronic lung disease suggests bronchiectasis or lung abscess
     • HIV or immunosuppression suggests neoplasia, TB, or Kaposi’s sarcoma
     • Tobacco use suggests bronchitis, lung cancer, or pneumonia
     • Weight loss suggests emphysema, lung cancer, tuberculosis, bronchiectasis, lung abscess, or HIV
• See Table 68.1

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infectious</td>
<td>Bronchitis, pneumonia (viral, bacterial, fungal), bronchiectasis, aspergillosis, tuberculosis</td>
</tr>
<tr>
<td>Malignancy</td>
<td>Primary lung cancer or lung metastases</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>PE, pulmonary artery rupture, CHF, mitral stenosis</td>
</tr>
<tr>
<td>Vasculitis</td>
<td>Granulomatosis with polyangiitis (formerly known as Wegener’s granulomatosis) or Goodpasture’s syndrome</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>Chest trauma, foreign body, anticoagulation, epistaxis, bronchovascular fistula, pulmonary arteriovenous malformation</td>
</tr>
</tbody>
</table>


4. EVALUATION OF HEMOPTYSIS

• Labs: CBC, chemistry panel, PT, PTT, urinalysis, and oximetry
• Chest X-ray and high-resolution CT scan of the chest (tracheal or proximal bronchial lesions missed by CXR)
• Tests to consider: sputum culture, PPD, sputum for AFB, coccidioidomycosis titer, and antineutrophil cytoplasmic and antiglomerular basement membrane antibodies
• Fiberoptic bronchoscopy indicated for unexplained hemoptysis

5. MANAGEMENT OF MASSIVE HEMOPTYSIS

• See Figure 68.1
• Intubation (preferably selective intubation of normal lung) and mechanical ventilation with the affected lung kept dependent in the lateral decubitus position
• Transfuse platelets or fresh frozen plasma for thrombocytopenia or a coagulopathy
• Pulmonary angiogram and selective embolization of the bronchial artery
• Lobectomy or pneumonectomy in refractory cases
6. **COMPLICATIONS**

- Exsanguination
- Hemorrhagic shock
- Asphyxiation
- Respiratory failure

**REFERENCES**

CHAPTER 69
EVALUATION OF PLEURAL EFFUSIONS

1. EPIDEMIOLOGY
   - United States: 1.5 million cases annually; mostly due to CHF, bacterial PNA, malignancy, and PE
   - Worldwide: 320/100,000

2. PATHOPHYSIOLOGY
   - Increased fluid entering pleural space (from interstitium, capillaries in parietal pleura, or holes in diaphragm) or decreased drainage of pleural space by lymphatics in parietal pleura leads to effusion
   - Transudate: due to systemic process
     - Left-sided CHF: fluid accumulates in lung interstitium faster than the ability of lymphatics to drain pleural space, leading to bilateral effusion
     - Hepatic hydrothorax: cirrhotic ascites enter pleural space via holes in diaphragm, which leads to right-sided (90%) or bilateral (10%) effusion
   - Exudate: due to local process
     - Bacterial pneumonia: parapneumonic effusion or empyema (frankly purulent)
     - Cancer: most commonly lung, breast, lymphoma; also mesothelioma
     - Pulmonary embolism
     - Viral infection
     - Tuberculosis: hypersensitivity reaction to TB antigen
     - Chylothorax: damaged thoracic duct from trauma or mediastinal tumors
     - Hemothorax: trauma or malignancy

3. CLINICAL PRESENTATION
   - Dyspnea, cough, and pleuritic chest pain are common
   - Historical clues
     - Trauma history suggests hemothorax
     - Cancer history suggests malignant effusion
     - Recent abdominal surgical procedures suggest postoperative effusion, subphrenic abscess, pulmonary embolism
     - Alcohol abuse or pancreatic disease suggests pancreatic effusion
     - Chronic hemodialysis suggests heart failure or uremic pleuritis
     - Cirrhosis suggests hepatic hydrothorax, spontaneous bacterial empyema
     - Cardiac surgery suggests Dressler’s syndrome
     - Esophageal procedure suggests esophageal perforation
     - Asbestos exposure suggests mesothelioma, benign asbestos effusion
     - Childbirth suggests postpartum pleural effusion
     - HIV infection suggests pneumonia, TB, lymphoma, Kaposi’s sarcoma
     - Rheumatoid arthritis suggests rheumatoid pleuritis
     - Lupus suggests lupus pleuritis, pneumonia, pulmonary embolism
   - Signs
     - Ascites: hepatic hydrothorax, ovarian cancer, Meigs’ syndrome
     - Dyspnea on exertion, orthopnea, peripheral edema: CHF
     - Pericardial friction rub, pericarditis
     - Unilateral leg swelling: PE
     - Yellowish nails, lymphedema: yellow nail syndrome
     - Fever: pneumonia, empyema, tuberculosis
Evaluation of Pleural Effusions

- See Tables 69.1 through 69.3

### TABLE 69.1. Classification of Pleural Effusions

<table>
<thead>
<tr>
<th>Test</th>
<th>Transudate</th>
<th>Exudate*</th>
</tr>
</thead>
<tbody>
<tr>
<td>PF NT-proBNP†</td>
<td>1300 pg/mL or greater</td>
<td>Less than 1300 pg/mL</td>
</tr>
<tr>
<td>Serum-PF albumin gradient†</td>
<td>Greater than 1.2 g/dL</td>
<td>Up to 1.2 g/dL</td>
</tr>
<tr>
<td>Serum-PF protein gradient†</td>
<td>3.1 mg/dL or greater</td>
<td>Less than 3.1 mg/dL</td>
</tr>
<tr>
<td>PF protein/serum protein</td>
<td>Up to 0.5</td>
<td>Greater than 0.5</td>
</tr>
<tr>
<td>PF LDH (international units)</td>
<td>Up to 200</td>
<td>Greater than ( \frac{2}{3} ) upper limit of labs normal range (or greater than 200)</td>
</tr>
<tr>
<td>PF LDH/serum LDH</td>
<td>Up to 0.6</td>
<td>Greater than 0.6</td>
</tr>
</tbody>
</table>

PF = pleural fluid, NT = N-terminal (as in NT-pro-BNP), BNP = B-type natriuretic peptide, LDH = lactate dehydrogenase
* Light’s criteria: only one test needs to be abnormal to classify effusion as an exudate; 80–85% accurate for exudates:
  + PF glucose less than 60 suggests cancer, tuberculosis, empyema, or effusion from lupus or RA
  + Bloody effusion suggests CA, tuberculosis, PE, or trauma
  + PF lymphocytosis greater than 50%: 90–96% from CA or tuberculosis
  + PF pH less than 7.2: empyema, malignancy, RA, or SLE
† Useful to diagnose transudative effusions after patient has received diuretics


### TABLE 69.2. Causes of Transudative Effusions

- Constrictive pericarditis
- Urinothorax
- Hepatic hydrothorax
- Heart failure*  
- Nephrotic syndrome
- Peritoneal dialysis
- Severe hypoalbuminemia*
- Superior vena cava syndrome

* Most common causes of transudative pleural effusions

4. COMPLICATIONS

- Trapped lung (formation of a restrictive, fibrous pleural peel around the visceral pleura)
- Empyema
- Severe sepsis

REFERENCES

CHAPTER 70
ASTHMA EXACERBATION

1. EPIDEMIOLOGY
   • Prevalence in United States is around 8% of population (9% for kids younger than 18 yo)
   • Mortality is 2/100,000/year
   • Most common reason for exacerbation is inadequate medical therapy combined with noncompliance
   • Predominantly in boys during childhood: male-to-female ratio of 2:1 until puberty, then it becomes 1:1; 50% of these children have symptom resolution by early adulthood; boys are more likely to have symptom resolution than girls
   • Most adult-onset asthma is diagnosed over age 40 yo in women
   • United States spends about 14 billion dollars per year on asthma, about 25% of which is on asthma exacerbations

2. PATHOPHYSIOLOGY
   • Exposure to allergens causes inflammation driven by mast cells, dendritic cells, and eosinophils, which activate inflammatory mediators leading to simultaneous inflammation and repair in airways
   • Epithelial damage
   • Subepithelial fibrosis results in basement membrane thickening
   • Smooth muscle hypertrophy results in airway hyperresponsiveness, which enables reversible obstruction
   • Increased vascular flow leads to airway edema
   • Goblet cell and submucosal gland hypertrophy → mucus hypersecretion

3. CLINICAL PRESENTATION
   • Signs: use of accessory muscles, chest wall retractions, tachypnea, cyanosis, hypoxia
   • Mild: dyspnea with activity
   • Moderate: dyspnea that prevents usual activity
   • Severe: dyspnea at rest, interferes with conversation

4. DIAGNOSIS
   • Reversible airflow obstruction on spirometry confirms asthma diagnosis
     ◦ FEV₁ less than 80% predicted or FEV₁/FVC less than 70%
     ◦ FEV₁ increases to 12% or more and FVC increases to 200 mL or FVC increases to 12% or more after inhaled β₂-agonist
     ◦ Bronchoprovocation test with inhaled methacholine can diagnose hyperbronchial responsiveness, which is suggestive of asthma
       ▪ Consider this test if spirometry is normal and clinical history very suggestive of asthma

5. DIFFERENTIAL DIAGNOSIS
   • COPD
   • Churg-Strauss syndrome
   • Eosinophilic pneumonia
   • CHF
   • Vocal cord paralysis
   • Vocal cord dysfunction
   • Foreign body aspiration
   • Laryngotracheal masses
- Tracheomalacia
- Angioedema
- Bronchiectasis
- Allergic bronchopulmonary aspergillosis
- Cystic fibrosis
- Bronchiolitis obliterans
- Conversion disorders
- Munchausen syndrome
- Malingering

6. EVALUATION

- Pulmonary function testing
- Consider allergen skin testing if exogenous trigger suspected
- See Tables 70.1 through 70.3

TABLE 70.1. Classification of Asthma Severity*

<table>
<thead>
<tr>
<th>Class</th>
<th>Days with Symptoms</th>
<th>Nights with Symptoms</th>
<th>PEF or FEV$^\dagger$</th>
<th>SABA Use for Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild intermittent</td>
<td>2 or fewer/week</td>
<td>2 or fewer/month</td>
<td>80% or more</td>
<td>2 days or fewer/week</td>
</tr>
<tr>
<td>Mild persistent</td>
<td>3–6/week</td>
<td>3–4/month</td>
<td>80% or more</td>
<td>More than 2 days/week</td>
</tr>
<tr>
<td>Moderate persistent</td>
<td>Daily</td>
<td>More than 1/week</td>
<td>More than 60% up to 80%</td>
<td>Daily</td>
</tr>
<tr>
<td>Severe persistent</td>
<td>Daily</td>
<td>Most nights</td>
<td>Up to 60%</td>
<td>Several times/day</td>
</tr>
</tbody>
</table>

PEF = peak expiratory flow, variability = daily variability over 1–2 weeks, FEV$1$ = Forced expiratory volume in 1 second, SABA = short-acting β$\_2$-agonist
* Same criteria used for children under 5 although spirometry not possible
† % personal best for PEF, % predicted for FEV$1$; may not correlate with symptoms


TABLE 70.2. Predicted PEF (Liters/Min) for Nonsmoking Patients

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Women (height in inches)</th>
<th>Men (height in inches)</th>
<th>Children</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>55</td>
<td>60</td>
<td>65</td>
</tr>
<tr>
<td>20</td>
<td>390</td>
<td>423</td>
<td>460</td>
</tr>
<tr>
<td>30</td>
<td>380</td>
<td>413</td>
<td>448</td>
</tr>
<tr>
<td>40</td>
<td>370</td>
<td>402</td>
<td>436</td>
</tr>
<tr>
<td>50</td>
<td>360</td>
<td>391</td>
<td>424</td>
</tr>
<tr>
<td>60</td>
<td>350</td>
<td>380</td>
<td>412</td>
</tr>
<tr>
<td>70</td>
<td>340</td>
<td>369</td>
<td>400</td>
</tr>
</tbody>
</table>

Source: Adapted from Am Rev Resp Dis, 1963; 88: 644–51.
TABLE 70.3. Risk Factors for Death in Asthmatics

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sudden, severe attacks</td>
<td>Prior intubation/ICU stay</td>
</tr>
<tr>
<td>Hospital/ER in last month</td>
<td>Recent systemic steroids</td>
</tr>
<tr>
<td>Cardiac problems</td>
<td>Illicit drug use</td>
</tr>
<tr>
<td>Psychosocial problems</td>
<td>Lack of asthma action plan</td>
</tr>
</tbody>
</table>


7. TREATMENT

Trigger Avoidance/Control
- Possible triggers: smoke, allergens, medications (β blocker, aspirin, NSAIDs)
- Exercise-induced: starts during and peaks 5–10 minutes after exercise
  - Inhaled β₂ agonist or mast cell stabilizer for prophylaxis
- Allergic rhinitis: control with intranasal steroids, allergen avoidance
- Gastroesophageal reflux: raise head of bed, avoid bedtime snack, medications

Stepwise Approach to Stable Asthma Management
- Gain control early with oral steroids or high-dose inhaled steroids
- Step down therapy every 1–2 months to least medications necessary
- Never use salmeterol or formoterol alone without an inhaled steroid
- Consider anti-IgE therapy if severe allergic asthma with elevated serum IgE
- See Tables 70.4 and 70.5 and Figure 70.1

TABLE 70.4. Management of Stable Asthma

<table>
<thead>
<tr>
<th>Class</th>
<th>Preferred Meds</th>
<th>Additional Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild intermittent</td>
<td>SA β₂-agonist* prn</td>
<td>--</td>
</tr>
<tr>
<td>Mild persistent</td>
<td>Low-dose inhaled steroids (see Table 70.5) and SA β₂-agonist* prn</td>
<td>Mast cell stabilizers (children) or leukotriene receptor blockers</td>
</tr>
<tr>
<td>Moderate persistent</td>
<td>LA β₂-agonists and low–medium dose inhaled steroids</td>
<td>Leukotriene receptor blockers or zileuton with or without theophylline SR‡</td>
</tr>
<tr>
<td>Severe persistent</td>
<td>High dose inhaled steroids (See Table 70.5) AND LA β₂-agonists†</td>
<td>Oral steroids with or without omalizumab if elevated serum IgE level</td>
</tr>
</tbody>
</table>

* SA = short-acting β₂-agonists: albuterol and levalbuterol used for breakthrough symptoms in all classes
† LA = long-acting β₂-agonists: salmeterol and formoterol
‡ Theophylline SR titrated to level 5–15 mcg/mL
TABLE 70.5. Inhaled Steroids: Recommended Daily Doses for Adults*

<table>
<thead>
<tr>
<th>Drug</th>
<th>Form</th>
<th>Adult Daily Doses</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Low</td>
</tr>
<tr>
<td>Beclomethasone MDI</td>
<td>40 mcg/puff</td>
<td>2–6</td>
</tr>
<tr>
<td></td>
<td>80 mcg/puff</td>
<td>1–3</td>
</tr>
<tr>
<td>Budesonide DPI</td>
<td>180 mcg/dose</td>
<td>1–3</td>
</tr>
<tr>
<td></td>
<td>Soln for nebs</td>
<td>–</td>
</tr>
<tr>
<td>Ciclesonide MDI</td>
<td>80 mcg/puff</td>
<td>1–3</td>
</tr>
<tr>
<td></td>
<td>160 mcg/puff</td>
<td>1–2</td>
</tr>
<tr>
<td>Flunisolide MDI</td>
<td>250 mcg/puff</td>
<td>2–4</td>
</tr>
<tr>
<td></td>
<td>80 mcg/puff</td>
<td>2–6</td>
</tr>
<tr>
<td>Fluticasone MDI</td>
<td>110 mcg/puff</td>
<td>1–2</td>
</tr>
<tr>
<td></td>
<td>220 mcg/puff</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>44 mcg/dose</td>
<td>2–6</td>
</tr>
<tr>
<td>Fluticasone DPI</td>
<td>100 mcg/dose</td>
<td>1–3</td>
</tr>
<tr>
<td></td>
<td>250 mcg/dose</td>
<td>1</td>
</tr>
<tr>
<td>Mometasone DPI</td>
<td>220 mcg/dose</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>110 mcg/dose</td>
<td>1–2</td>
</tr>
<tr>
<td>Triamcinolone MDI</td>
<td>75 mcg/puff</td>
<td>4–8</td>
</tr>
</tbody>
</table>

* MDI = metered dose inhaler, DPI = dry powder inhaler; all doses in puffs (MDI) or inhalations (DPI)

8. COMPLICATIONS

- Pneumothorax
- Respiratory failure

REFERENCES


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**Initial Assessment of Severity in Acute Asthma Exacerbations in Adults**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speaking in</td>
<td>Sentences</td>
<td>Phrases</td>
<td>Words</td>
</tr>
<tr>
<td>Heart rate</td>
<td>Less than 100 beats/min</td>
<td>100–120 beats/min</td>
<td>More than 120 beats/min</td>
</tr>
<tr>
<td>PEF/FEV₁ (% predicted)</td>
<td>Greater than 70%</td>
<td>40–70%</td>
<td>Less than 40% (esp. Less than 25%)</td>
</tr>
<tr>
<td>Room air pulse oximetry</td>
<td>Greater than 95%</td>
<td>91–95%</td>
<td>or less 90%</td>
</tr>
<tr>
<td>Mental status</td>
<td>Alert</td>
<td>Drowsy</td>
<td>Lethargic/obtunded</td>
</tr>
<tr>
<td>PaCO₂ (mmHg)</td>
<td>Less than 40</td>
<td>40–90</td>
<td>Greater than 50t</td>
</tr>
</tbody>
</table>

**Inpatient treatment of moderate–severe asthma exacerbations**

- Oxygen to keep SaO₂ above 90%
- Albuterol 2.5 mg Neb q1 h until stable, then q 2 h/q 1 h prn x 24 h, then q 4 h/q 2 h prn
- Consider 2.5 mg Neb q 20 min x 3 or 10 mg continuous over 1 h for severe asthma
- Albuterol and levalbuterol are equally efficacious
- Ipratropium 0.5 mg Neb q 20 min x 3 for severe asthma, then q 4 h/q 2 h prn, then q 6 h
- Methylprednisolone 60 mg IV q 6 h until bronchospasm controlled, then prednisone 1 mg/kg PO daily to complete 10–14 days of therapy, then taper
- Consider magnesium 2 g IV over 20 minutes for severe asthma exacerbations
- Empiric antibiotics for pneumonia or bronchitis only if purulent sputum production

**Immediate treatment**

- Serial PEF and oximetry monitoring
- Smoking cessation counseling, if applicable, and patient education

**Good response after 1–2 h**

- Continue current therapy on wards
- Serial PEF and oximetry monitoring
- Smoking cessation counseling, if applicable, and patient education

**Partial/poor response after 1–2 h**

- Consider noninvasive positive pressure ventilation in ICU
- Continue q 1 h nebulizer treatments

**Reassess after 1–2 h**

- ABG
- Serial PEF and oximetry monitoring
- Consider IV aminophylline only for severe, refractory asthma (high risk of toxicity)
- Consider intubation for persistent PaCO₂ Greater than 50 with respiratory acidosis, worsening mental status, hemodynamic instability, or progressive deterioration despite maximal medical therapy
- Propofol and ketamine will bronchodilate
- Maximize expiratory time
- Keep plateau at 30 cmH₂O

**FIGURE 70.1. Management of Acute Asthma Exacerbations**

CHAPTER 71

CHRONIC OBSTRUCTIVE PULMONARY DISEASE EXACERBATION

1. EPIDEMIOLOGY
   - Fourth leading cause of death in the United States
   - Prevalence is 20% in the United States
   - Cigarette smoking is implicated in 90% of cases
   - 75% of patients have serious chronic dyspnea and nearly 25% have profound total body pain
   - 60 yo smoker with chronic bronchitis has a 10-year mortality of 60%, which is 4 times higher than age-matched nonsmoking asthmatics
   - Inpatient mortality is 11%, 6-month mortality is 33%, and 1-year mortality is 43%
   - Those who survived first hospitalization have a 50% chance of rehospitalization within 6 months
   - Initial hospitalization: 93% are male, mean age is 63.5 yo, less than 1% never smoked, mean FEV₁ is 47%, and 50% are admitted to the ICU

2. PATHOPHYSIOLOGY
   - Large airways: mucus and goblet cell hyperplasia increase mucus, which leads to cough then chronic bronchitis
   - Small airways: irreversible airway obstruction (decreased FEV₁) with compensatory hyperinflation (increased residual volume)
   - Initially, air trapping maintains airflow (increased lung volume increases elastic recoil, increased airway diameter decreases airway resistance)
   - Hyperinflation flattens diaphragm decreases inspiratory capacity
   - Decreased abdominal pressure transmitted to diaphragm
   - Shorter, less effective diaphragmatic muscle fibers
   - Due to Laplace's law, need to increase tension to produce a given pressure
   - Lung parenchyma: with chronic inflammation, elastase activity exceeds antielastase activity, leading to degradation of extracellular matrix, cell death, and patchy enlarged air spaces (i.e., emphysema)
     - FEV₁ less than 50% predicted associated with hypoxemia
     - FEV₁ less than 25% predicted associated with hypercapnia

3. CLINICAL PRESENTATION
   - Acute change from baseline dyspnea, cough, or sputum production
   - Other symptoms: chest tightness; tachycardia; decreased exercise tolerance; confusion; depression; insomnia; change to color, volume, or tenacity of sputum; dyspnea; tachypnea; wheezing; fever; fatigue; malaise
   - Physical findings
     - Cardiac impulse palpable below the xiphoid [LR+ 7.4, LR– NS]
     - Hoover sign: hands placed on costal margin, with fingers touching at xiphoid process—with normal respiration, the hands will separate; in COPD, the hyperexpansion prevents further excursion and the hands come closer together [LR+ 4.2, LR– 0.5]
     - Accessory (scalene/sternocleidomastoid) muscle use [LR+ 3.3, LR– 0.7]
     - Decreased breath sounds [LR+ 3.2, LR– 0.5]
     - Wheeze [LR+ 2.8, LR– 0.8]
     - Barrel chest [LR+ 1.5, LR– 0.6]
4. **DIAGNOSIS**
   - Spirometry: airflow obstruction that is not fully reversible
     - FEV₁/FVC less than 70% predicted and postbronchodilator FEV₁ less than 80%
   - **Diagnosis of COPD exacerbations**: increase in dyspnea, cough, sputum volume, or purulence

5. **DIFFERENTIAL DIAGNOSIS**
   - Asthma
   - CHF
   - Angioedema
   - Bronchiectasis
   - Allergic bronchopulmonary aspergillosis
   - Cystic fibrosis
   - Bronchiolitis obliterans
   - Conversion disorders
   - Munchausen syndrome
   - Malingering
   - Interstitial lung disease

6. **EVALUATION**
   - Baseline pulmonary function test, pulse oximetry, chest X-ray
   - Labs: α₁-antitrypsin

**Evaluation of COPD Exacerbations**
- Assess with chest X-ray, sputum culture, oximetry, or arterial blood gas
- Admit for moderate–severe exacerbations: respiratory acidosis, need for ventilation, PEF less than 100 L/min, FEV₁ less than 1 L or less than 40% predicted, or serious comorbidities
- See Table 71.1

<table>
<thead>
<tr>
<th>TABLE 71.1. Management of Stable COPD by Stage</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stage</strong></td>
</tr>
<tr>
<td>All (at risk)</td>
</tr>
<tr>
<td>I (mild)</td>
</tr>
<tr>
<td>II (moderate)</td>
</tr>
<tr>
<td>III (severe)</td>
</tr>
<tr>
<td>IV (very severe)</td>
</tr>
</tbody>
</table>

* FEV₁ used to stratify severity

BODE index (body mass index, airway obstruction, dyspnea, exercise capacity on 6 min. walk) better to assess risk of death: see http://content.nejm.org/cgi/reprint/350/10/1005.pdf

† Bronchodilators (anticholinergics > β₂-agonists > methylxanthines): use combination therapy if monotherapy inadequate; long-acting anticholinergic (tiotropium) and β₂-agonist (e.g., salmeterol or formoterol) are preferred over short-acting anticholinergic (praprovonium) or β₂-agonist (e.g., albuterol)

‡ Aerobic exercise, good nutrition and education

§ PaO₂ < 55 mmHg/SpO₂ < 88% (PaO₂ < 60 mmHg if pulmonary hypertension, polycythemia, or cor pulmonale)

|| Bullectomy or lung-volume reduction surgery best for upper lobe emphysema and low exercise capacity; lung transplantation indicated for idiopathic emphysema or α₁-antitrypsin deficiency

¶ Roflumilast 500 mcg PO daily (PDE4 inhibitor) decreases COPD exacerbations

7. **TREATMENT OF ACUTE COPD EXACERBATIONS**

- Albuterol 2.5 mg and ipratropium 0.5 mg nebulized q 2–4 h
- Antibiotics × 5–10 days for severe exacerbations or presence of purulent sputum
  - Uncomplicated exacerbation if age 65 yo or younger, FEV1 50% or more, and fewer than 4 exacerbations/year
    - New macrolide, doxycycline, or 2nd–3rd generation cephalosporin
  - Complicated exacerbation if age older than 65 yo, FEV1 less than 50%, more than 4 exacerbations/year, or use of antibiotics in the last 3 months
    - Use amoxicillin-clavulanate or a respiratory quinolone
- Risk for pseudomonas if recurrent antibiotic use, recurrent steroid courses, or if bronchiectasis is present; use an antipseudomonal quinolone
- Systemic steroids with methylprednisolone 30–40 mg/day (or prednisone 40–60 mg PO daily) × 7–10 days if FEV1 is less than 50% predicted, or if PaCO2 is greater than 45 and pH is less than 7.35 with or without steroid taper
- Oxygen if hypoxia to maintain SaO2 is 88–90% or PaO2 is 55 mmHg or higher
- Noninvasive positive-pressure ventilation if acute respiratory acidosis (pH 7.35 or less, PaCO2 45 mmHg or higher) and no contraindications to its use
- Indications for ICU admission: PaCO2 greater than 60 mmHg and pH less than 7.25, depressed level of consciousness, unstable medical comorbidities, hemodynamic or rhythm instability, and need for invasive mechanical ventilation
- Indications for mechanical ventilation: severe respiratory acidosis refractory to noninvasive ventilation, respiratory arrest, hemodynamic instability, or obtundation

8. **COMPLICATIONS**

- Progressive dyspnea
- Respiratory failure
- Frequent/recurrent pulmonary infections
- Pulmonary hypertension results in cor pulmonale
- Depression

**REFERENCES**

CHAPTER 72
PERIOPERATIVE PULMONARY EVALUATION AND MANAGEMENT

1. PREOPERATIVE RISK STRATIFICATION
   - Few patients have an absolute pulmonary contraindication to surgery
   - Preoperative spirometry should not be used to prevent surgery but rather as a tool to optimize preoperative lung function; appropriate if the patient has
     - Asthma or COPD and air flow obstruction that has not been optimized
     - Unexplained dyspnea or cough and will undergo major surgery (as below)
     - Patient will be undergoing lung resection surgery
   - Indications for an arterial blood gas: resting hypoxia, risk for chronic hypercapnia, or anticipated lung resection surgery

2. PERIOPERATIVE PULMONARY MANAGEMENT TABLES

   **TABLE 72.1. Risk Factors for Perioperative Pulmonary Complications**

<table>
<thead>
<tr>
<th>Factor</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age older than 60 years</td>
<td></td>
</tr>
<tr>
<td>Smoking within 8 weeks of surgery</td>
<td></td>
</tr>
<tr>
<td>Poor general health (ASA 3 or more)†</td>
<td></td>
</tr>
<tr>
<td>Emergency surgery</td>
<td></td>
</tr>
<tr>
<td>Thoracic, abdominal aortic aneurysm, neurosurgery, head and neck or upper abdominal surgery</td>
<td></td>
</tr>
<tr>
<td>Surgery lasting more than 3 hours</td>
<td></td>
</tr>
<tr>
<td>General anesthesia</td>
<td></td>
</tr>
<tr>
<td>Long-acting neuromuscular blockade</td>
<td></td>
</tr>
<tr>
<td>COPD</td>
<td></td>
</tr>
<tr>
<td>CHF</td>
<td></td>
</tr>
<tr>
<td>Elevated arterial carbon dioxide pressure (PaCO₂ 45 mmHg)</td>
<td></td>
</tr>
<tr>
<td>Functional dependence for ADLs</td>
<td></td>
</tr>
<tr>
<td>Impaired sensorium</td>
<td></td>
</tr>
<tr>
<td>Malnourished (albumin less than 3.5 g/dL)</td>
<td></td>
</tr>
<tr>
<td>Renal failure (blood urea nitrogen [BUN] 21 mg/dL or higher)</td>
<td></td>
</tr>
<tr>
<td>Transfusion more than 4 units of blood</td>
<td></td>
</tr>
</tbody>
</table>

   Note: Obesity alone or asthma does not appear to increase risk

   ASA = aspirin, COPD = chronic obstructive pulmonary disease, CHF = congestive heart failure, ADL = activities of daily living, BUN = blood urea nitrogen

   † American Society of Anesthesiologists Classification at www.asahq.org/clinical/physicalstatus.htm


   **TABLE 72.2. Postoperative Respiratory Failure Index**

<table>
<thead>
<tr>
<th>Factor</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of surgery</td>
<td></td>
</tr>
<tr>
<td>Albumin less than 3 g/dL</td>
<td>9</td>
</tr>
<tr>
<td>AAA repair</td>
<td>27</td>
</tr>
<tr>
<td>BUN greater than 30 mg/dL</td>
<td>8</td>
</tr>
<tr>
<td>Thoracic surgery</td>
<td>21</td>
</tr>
<tr>
<td>History of COPD</td>
<td>6</td>
</tr>
<tr>
<td>Neurosurgery, upper abdomen, or peripheral vascular surgery</td>
<td>14</td>
</tr>
<tr>
<td>Partially or fully dependent functional status</td>
<td>7</td>
</tr>
<tr>
<td>Neck surgery</td>
<td>11</td>
</tr>
<tr>
<td>Age 70 yo or older</td>
<td>6</td>
</tr>
<tr>
<td>Emergency surgery</td>
<td>11</td>
</tr>
<tr>
<td>Age 60–69 yo</td>
<td>4</td>
</tr>
</tbody>
</table>

   **Class**  **Points**  **Incidence of Postoperative Respiratory Failure**

<table>
<thead>
<tr>
<th>Class</th>
<th>Points</th>
<th>Incidence of Postoperative Respiratory Failure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10 or fewer</td>
<td>0.5%</td>
</tr>
<tr>
<td>2</td>
<td>11–19</td>
<td>1.8%</td>
</tr>
<tr>
<td>3</td>
<td>20–27</td>
<td>4.2%</td>
</tr>
<tr>
<td>4</td>
<td>28–40</td>
<td>10.1%</td>
</tr>
<tr>
<td>5</td>
<td>More than 40</td>
<td>26.6%</td>
</tr>
</tbody>
</table>

   Source: Adapted from Ann Intern Med, 2006;144:575.
3. INTERVENTIONS TO REDUCE PERIOPERATIVE RISK

- Smoking cessation: beneficial if patient quits 8 or more weeks prior to surgery
- Inhaled tiotropium 1 puff daily for COPD with or without β₂ agonists for wheezing
- Oral or inhaled steroids and inhaled tiotropium if COPD or asthma and pulmonary function not optimal (no increase in risk of infections, but potential for adrenal suppression if 20 mg/day or more of prednisone for at least 3 weeks)
- Defer elective surgery for acute exacerbations of pulmonary disease
- Consider shorter procedures (under 3 hours), laparoscopic approach, and spinal/epidural or regional anesthesia rather than general anesthesia for high-risk patients
- Avoid long-acting neuromuscular blockers (e.g., pancuronium)
- Postoperative lung expansion maneuvers and early mobilization recommended
- Consider postoperative epidural analgesia for thoracic or upper abdominal surgery

REFERENCES

CHAPTER 73
DIFFUSE INTERSTITIAL LUNG DISEASE

1. EPIDEMIOLOGY
   - 75% of IPF are older than 60 yo at diagnosis
   - Almost all patients with lymphangioleiomyomatosis (LAM) are women

2. PATHOPHYSIOLOGY
   - Predominant histopathological patterns
     - Granulomatous: T cells, macrophages, and epitheloid cells organized into granulomas
       - Known cause: hypersensitivity pneumonitis
       - Unknown cause: sarcoidosis, granulomatosis with polyangiitis (formerly known as Wegener’s granulomatosis), Churg-Strauss
     - Inflammatory/fibrotic: epithelial injury induces alveolar inflammation; interstitial and vascular inflammation leads to interstitial fibrosis, irreversible scarring, and impaired gas exchange
       - Known cause: asbestos, inhalation, medications (nitrofurantoin, amiodarone), chemotherapy (bleomycin), radiation, aspiration, post-ARDS, desquamative interstitial pneumonia, Langerhans cell granulomatosis
       - Unknown cause: idiopathic pulmonary fibrosis (usual interstitial pneumonia), diffuse alveolar damage, cryptogenic organizing pneumonia, nonspecific interstitial pneumonia, rheumatologic diseases, Goodpasture’s syndrome, pulmonary alveolar proteinosis, eosinophilic pneumonia, lymphangioleiomyomatosis, amyloidosis, genetic diseases, graft-versus-host, etc.

3. CLINICAL PRESENTATION
   - Progressive dyspnea and cough
   - Acuity of onset ranges from years (UIP) to days/weeks (AIP)
   - Signs: crackles, inspiratory squeaks; rarely cor pulmonale

4. DIAGNOSIS
   - Definitive diagnosis is via a tissue biopsy
   - History: occupational/environmental exposures, travel, meds, medical comorbidities

5. DIFFERENTIAL DIAGNOSIS
   - CHF
   - Fungal pneumonia
   - Miliary tuberculosis
   - Pulmonary hypertension
   - Interstitial spread of cancer

6. EVALUATION
   - Chest X-ray and arterial blood gas
   - High-resolution chest CT: reticulonodular infiltrates, interstitial infiltrates, “ground glass” opacities, or honeycombing
   - Pulmonary function testing reveals a pattern of restrictive lung disease with a decline in lung volumes and diffusion capacity
   - Labs: CBCD, chemistry panel, angiotensin converting enzyme, ANA, RF, antineutrophil cytoplasmic antibody, antiglomerular basement membrane antibody, anti-Scl-70, HIV, ESR, CK, HLA-B27, aldolase levels, and coccidioidomycosis titers
   - Induced sputa or bronchoalveolar lavage for cytology and AFB or fungi
   - Open lung biopsy
   - See Figure 73.1
7. TREATMENT OF DIFFUSE INTERSTITIAL LUNG DISEASE (DILD)
   • Targeted therapy of any underlying disease that has been identified
   • Avoidance of offending medication(s) or occupational exposures if relevant
   • Supplemental O₂ to keep PaO₂ above 55 mmHg; pneumococcal and influenza vaccines
   • Idiopathic interstitial pneumonitis: prednisone 0.5–1 mg/kg/day often, with cyclophosphamide 2 mg/kg PO daily OR azathioprine 2 mg/kg PO daily
   • Refer appropriate patients for lung transplantation

8. COMPLICATIONS
   • Pulmonary hypertension
   • Cor pulmonale
   • Respiratory failure

REFERENCES
CHAPTER 74
PULMONARY HYPERTENSION

1. EPIDEMIOLOGY
   • Uncommon; incidence 2.4–7.6 cases per million, prevalence of 15–50 cases per million
   • Poor prognosis with 15% mortality within 1 year
   • Male to female ratio is 1:2; mean age of onset is around 37 yo; idiopathic type is most common at 39%, 70% arise from a germine mutation in BMPR2 gene
   • Drug-induced PAH is primarily related to anorexigen (diet pill), 10%; other drugs such as amphetamines and L tryptophan are also associated with its development
   • 25–50% of patients suffer from Eisenmenger syndrome
   • 4% or patients with PE go on to develop PAH
   • 4% PE patients go on to develop PAH

2. CLINICAL PRESENTATION
   • Early phases are asymptomatic
   • Presenting symptoms are dyspnea on exertion, fatigue, chest pain, palpitations, and edema
   • Signs: R parasternal lift, accentuated second heart sound, pansystolic murmur (tricuspid regurgitation), third heart sound, diastolic murmur (pulmonary valve insufficiency), peripheral edema
   • Risk factors are congenital heart disease, connective tissue disease, portal hypertension, sickle cell disease, thyroid disease, and HIV

3. DEFINITION
   • Mean pulmonary artery pressure is greater than 25 mmHg (rest)
   • Pulmonary arterial hypertension (PAH): above and normal PCWP or normal LVEDP
   • See Table 74.1

TABLE 74.1. Classification of Pulmonary Hypertension (PHTN)

<table>
<thead>
<tr>
<th>Subtypes of PHTN</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary arterial hypertension</td>
<td>• Idiopathic&lt;br&gt; • Familial&lt;br&gt; • Associated with a connective tissue disease&lt;br&gt; • Associated with HIV infection&lt;br&gt; • Portopulmonary hypertension&lt;br&gt; • Drug-induced or toxin-induced&lt;br&gt; • Pulmonary veno-occlusive disease&lt;br&gt; • Pulmonary capillary hemangiomatosis</td>
</tr>
<tr>
<td>with left heart disease</td>
<td>• Chronic CHF, moderate–severe MS or MR</td>
</tr>
<tr>
<td>Secondary to chronic hypoxia</td>
<td>• COPD, DILD, OSA, obesity-hypoventilation syncope, neuromuscular disorders, intracardiac right-to-left shunts</td>
</tr>
<tr>
<td>Recurrent pulmonary emboli</td>
<td>• Recurrent PE or tumor emboli</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>• Sarcoidosis, Langerhans cell histiocytosis, schistosomiasis, and lymphangiomatosis</td>
</tr>
</tbody>
</table>

Source: Adapted from J Am Coll Cardiol, 2004(12 Suppl S); 43: 55–12S.
4. EVALUATION OF PULMONARY HYPERTENSION

- ECG: right axis deviation, right atrial abnormality, and right ventricular hypertrophy
- Chest X-ray: enlarged right atrium and ventricle, dilated pulmonary arteries, and “pruning” of the peripheral pulmonary vasculature
- Pulmonary function test: screen for underlying obstructive or restrictive lung disease
- Nocturnal polysomnogram: evaluate for obstructive or central sleep apnea
- Arterial blood gas: screen for resting hypoxia or hypercarbia
- V/Q scan superior to CT pulmonary angiogram to screen for recurrent PE
- Echocardiogram: evaluate left ventricular systolic and diastolic function, valve abnormalities, chamber sizes, and noninvasive estimate of pulmonary pressures
- Right heart catheterization: confirms diagnosis of pulmonary arterial hypertension
- Labs: CBCD, chemistry panel, ANA, RF, anti-Scl-70, anticentromere, HIV, and ESR
- 6-minute walk test to determine functional capacity

5. TREATMENT

- Supplemental oxygen to keep SaO₂ 90% or more
- Interventions: influenza and pneumococcal vaccinations; smoking cessation; chronic anticoagulation; and avoid decongestants, pregnancy, NSAIDs, and air travel
- Consider cautious diuresis and/or digoxin therapy for right ventricular dysfunction
- Pulmonary vasoreactivity as determined by inhaled nitric oxide testing during right heart catheterization can benefit from nifedipine ER, amlodipine, or diltiazem therapy
- Initial medication therapy for functional classes 2–3 and good hemodynamic profile
  - Endothelin receptor antagonists: bosentan or ambrisentan
  - Phosphodiesterase-5 inhibitors: sildenafil
- Initial medication therapy for functional classes 3–4 with poor hemodynamic profile
  - Prostanoid therapy: epoprostenol IV or inhaled iloprost or treprostinil SQ or IV

6. COMPLICATIONS

- Cor pulmonale
- Hypoxemia
- Cardiac arrhythmias
- Hemoptysis

REFERENCES