Emphysema

Description/Etiology

Emphysema is a progressive and permanent distention of the alveoli in the lungs that leads to destruction of the alveolar walls and impairment in oxygen and carbon dioxide (CO₂) gas exchange. Oxygen diffusion into the blood is impaired by the resulting decrease in direct contact of alveolar surface area with the pulmonary capillaries, leading to hypoxemia. In later stages of emphysema, the ability to exhale carbon dioxide is impaired, leading to hypercapnia (i.e., increased carbon dioxide in arterial blood) and, ultimately, respiratory acidosis (i.e., arterial blood becomes acidic from accumulated CO₂). The pathogenesis of emphysema is currently unclear; currently, two hypotheses have been proposed by researchers that attempt to explain its development. One theory suggests that inflammation of the airways leads to its eventual disappearance prior to emphysema development. Another theory suggested that vascular endothelial apoptosis’ potential role in inflammation might be what leads to emphysema. The slightest trauma to the river levees of the lungs’ airways can lead to an impairment of their natural protective systems, which could potentially lead to emphysema. Emphysema can affect all of the lung tissue or concentrate in specific areas. The following three morphological types of emphysema are recognized:

- **Centrilobular emphysema** (also called centriacinar emphysema) begins in the bronchioles and spreads peripherally, predominantly affecting the upper half of the lungs; this is the most common form of emphysema and is associated with long-standing cigarette smoking.
- **Panacinar emphysema** predominantly affects the lower lobes and is associated with an inherited deficiency in alpha 1-antitrypsin (AAT).
- **Paraseptal emphysema**, which primarily involves the distal airway structures, alveolar ducts, and alveolar sacs, occurs in young adults and is often caused by pneumothorax.

Treatment for emphysema involves pharmacologic agents (e.g., bronchodilators and inhaled corticosteroids), supplemental oxygen (indicated if pulse oximetry drops below 88%), and surgery (e.g., lung transplantation [usually reserved for patients with limited life expectancy] and lung volume reduction [i.e., resection]; for more information, see Quick Lesson About ... Emphysema: Lung Volume Reduction Surgery). Except for lung transplantation, treatment can improve quality of life but cannot reverse lung damage. Smoking cessation and physical activity prevent further deterioration of pulmonary function and can help reduce the need for supplemental oxygen. All patients should be administered albuterol in cases of emergencies. Antibiotics should be administered to patients who have moderate to severe exacerbations due to bacterial infections, as indicated and ordered.

Facts and Figures

Emphysema affects ~ 18;1,000 persons in the United States. Emphysema is a component of chronic obstructive pulmonary disease (COPD), a disease state that affects up to 10% of the population worldwide and is the third leading cause of mortality in the U.S (for more information on COPD, see Food for Thought, below). Between 80% and 90% of patients with COPD have a history of smoking, but only 15–20% of smokers develop COPD.

Risk Factors

Smoking is the primary risk factor for emphysema. Other risk factors for emphysema include prolonged and intense exposure to occupational dusts, chemicals, and air pollution;
intravenous drug use; and immunodeficiency syndromes (e.g., HIV infection/AIDS). AAT deficiency is inherited in an autosomal recessive pattern; the risk of inheriting AAT deficiency is 25% if both parents carry the causative gene mutation.

**Signs and Symptoms/Clinical Presentation**

The initial symptom of emphysema is usually dyspnea. Coughing, wheezing, and expectoration can occur, especially if bronchitis is also present, but emphysema can occur without cough. Persons with emphysema typically have adequate oxygen saturation and are referred to as “pink puffers.” They breathe through pursed lips and use accessory respiratory muscles as they try to empty CO$_2$ from the lungs. Barrel chest can occur as the lungs overinflate, and the patient might sit in a forward and hunched position to relieve chest pressure. Weight loss can occur as eating becomes difficult. Death is from respiratory acidosis and coma, heart failure, or lung collapse.

**Assessment**

› **Patient History**
  - Ask about history of smoking or exposure to secondhand smoke or other irritants, including occupational exposure; history of respiratory infections, especially bronchitis; family history of AAT deficiency; and recent weight loss, depression, or anxiety

› **Physical Findings of Particular Interest**
  - Auscultation of breath sounds will identify areas of involvement (e.g., bilateral or unilateral; upper, middle, or lower lobe)
  - Pursed lip breathing, hunched forward position, barrel chest, dyspnea, coughing, wheezing, and elevated heart rate are commonly present

› **Laboratory Tests That Can Be Ordered**
  - ABGs can indicate hypoxemia, ↑ CO$_2$, and compensated respiratory acidosis
  - Genetic testing for counseling on reproductive options is indicated if symptom onset is before age 50, particularly in a nonsmoker with family history of emphysema and/or liver disease

› **Other Diagnostic Tests/Studies**
  - EKG might indicate heart disease, which often occurs with emphysema
  - Frontal and lateral X-ray can show lung hyperinflation
  - High-resolution CT scan can detect areas of low attenuation from normal lung parenchyma and upper lobe foci most consistent with smoking-induced disease
    - In a study on the use of CT in detection of emphysema, researchers reported ultralow-dose chest CT performed for lung cancer can also help in detecting emphysema (Wang et al., 2015)
  - Pulmonary function tests can detect increased lung volume, air-trapping, decreased expiratory airflow, and destruction of capillary beds

**Treatment Goals**

› **Promote Optimum Respiratory Function and Reduce Risk of Complications**
  - Monitor vital signs, assess all physiologic systems (especially pulmonary), and review lab/diagnostic study results; immediately report abnormalities and treat, as ordered, including administering supplemental oxygen
  - Maintain patient safety (e.g., airway, circulation, and prevention of injury)
  - Administer prescribed pharmacologic agents, including bronchodilators (e.g., beta-2 agonists and/or anticholinergics), oral corticosteroids or other anti-inflammatories, and antibiotics (compromised lung function increases vulnerability to respiratory infection)
  - Follow facility pre- and postsurgical protocols if patient becomes a surgical candidate; reinforce pre- and postsurgical education and verify completion of facility informed consent documents
    - Postsurgically, assess for pain and administer prescribed analgesia
    - Frequently assess surgical site of giant bullae resection or lung volume reduction for infection, bleeding, or swelling; change dressings, as ordered
    - Following transplantation, administer anti-rejection drugs and assess for signs of infection, organ rejection, and graft-versus-host disease

› **Promote Emotional Well-Being and Educate**
  - Assess patient’s anxiety level and coping ability; educate and encourage discussion about emphysema etiology, treatment risks and benefits, potential complications, and individualized prognosis
• Reinforce the treating clinician’s recommendations regarding physical activity; depending on the patient’s lung function and overall health status, appropriate exercises may include walking, bicycling, and weight training

**Food for Thought**

› COPD has historically been described as encompassing emphysema and chronic bronchitis (i.e., inflammation of the bronchi and excessive mucous production in the trachea and bronchi resulting in the presence of a daily productive cough for 3 months or more each year for 2 consecutive years; see *Quick Lesson About ... Bronchitis, Chronic*). However, the current Global Initiative for Chronic Obstructive Lung Disease (GOLD) definition of COPD does not include these terms, instead defining COPD as “characterized by persistent airflow limitation that is usually progressive and associated with enhanced chronic inflammatory response in the airways and the lung to noxious particles or gases.” Because the onset, presence, severity, and contribution to disease status of chronic bronchitis and emphysema vary widely among persons with COPD, the definition of COPD in the GOLD report no longer includes these conditions as being definitive of COPD, but describes them as a variable part of the pathology of COPD

› Emphysema and pulmonary fibrosis, which have dissimilar physiologic effects (e.g., increased lung compliance and increased lung volumes in the former and decreased lung compliance and reduced lung volumes in the latter), are being increasingly recognized as coexisting in the same patient, leading to the recognition of a clinical syndrome called “combined pulmonary fibrosis and emphysema (CPFE) syndrome.” This syndrome, which primarily affects male smokers, is characterized by dyspnea, upper lobe emphysema, lower-lobe fibrosis, and impaired gas exchange

• Researchers recently found that CPFE is present in 8% of patients with idiopathic pulmonary fibrosis (IPF); mortality rates were similar in patients with comorbid IPF and emphysema and those with IPF without emphysema (Reyerson et al., 2013)

• Another research team found that a significant number of patients with CPFE have underlying autoimmune disorders and that these patients have a more favorable prognosis than those without evidence of autoimmunity (Tzouvelekis et al., 2013)

› Researchers described a case study involving a 17-year-old male patient who had subcutaneous emphysema, pneumomediastinum, and pneumo-orbita due to facial trauma following an accident. The researchers reported that the subcutaneous emphysema had extended to the mediastinal cavity, but no recorded incidents to date had involved a patient developing pneumomediastinum due to trauma. The trauma was caused by a pressurized car washer that impacted the patient’s face, leading to the aforementioned conditions (Yilmaz et al., 2014)

› Researchers involved in a recent randomized control trial found that Zephyr endobronchial valve (EBV) treatment displayed notable improvements in patients with severe heterogeneous emphysema. Patients were reported to have an improvement in lung function and overall improved quality of life due to decreased dyspnea. The prospective study included a total of 97 patients with emphysema of whom 55.4% receiving EBV had improved forced expiratory volume at 3 months; within 6 months, the number increased to 56.3%. Overall, the researchers concluded that EBV treatment has a good safety profile and can be used to treat symptoms of emphysema (Kemp et al., 2017)

› Researchers conducted a prospective study of 52 patients with COPD and found no increased risk of osteoporosis due to emphysema, despite initial hypotheses that had previously suggested a correlation. No observable differences were reported between patients with emphysema and patients without emphysema in regard to vertebral fracture scores, serum vitamin D levels, or bone mass density (Fouda et al., 2017)

**Red Flags**

› Potential complications of emphysema include cor pulmonale (i.e., damage to the right ventricle of the heart in response to resistance of the lungs), right ventricular hypertrophy or heart failure, depression over loss of independence with activities of daily living, and panic attacks associated with dyspnea

› Emphysema elevates the risk for certain bacterial and fungal lung infections

› Episodes of right-sided heart failure, peripheral edema, respiratory failure, and cyanosis can occur in patients with centrilobular emphysema

**What Do I Need to Tell the Patient/Patient’s Family?**

› Avoid any exposure to irritants (especially cigarette smoke) and minimize risk for respiratory infections through influenza and pneumococcal vaccinations, avoiding crowds, and diligent hand hygiene; adherence to oxygen therapy can extend life

› Exercise regularly, maintain a healthy diet, and take medications as prescribed, and notify the treating clinician or call 9-1-1 if new or worsening signs and symptoms (e.g., increased difficulty breathing, racing heart, trouble talking or walking) develop

› Disease progression can be slowed, but emphysema cannot be cured or reversed
References


