

## Emphysema – Causes and Treatment

[See online here](#)

**Emphysema is a condition characterized by the dilation of air spaces, with decreased elasticity and increased compliance, due to alveolar wall destruction; capillaries are also destroyed along with the alveoli. The most common cause of developing emphysema is smoking. Alpha-1-antitrypsin deficiency may also result in the development of emphysema.**



### Definition and Background of Emphysema

Emphysema is defined as an **unusual enlargement of the airspace** that is permanent and **distal to the terminal bronchioles**, which is combined with the destruction of the airspace walls, **without obvious fibrosis**.

Normal bronchioles and alveoli look like a cluster of grapes with each alveoli being divided by septae. However, if you have emphysema, you **lose your surface area**, which means you **do not have an effective gas exchange**; hence, your diffusing capacity of the [lungs](#) for carbon monoxide (DLCO) is reduced.

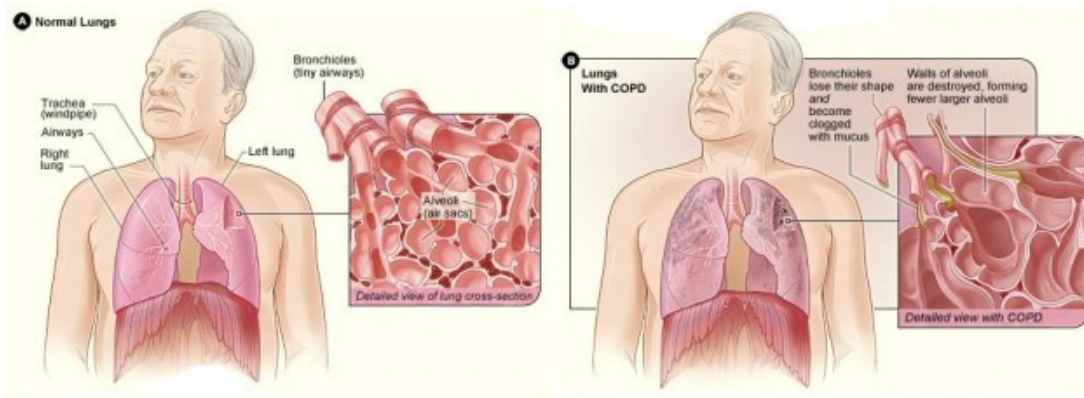


Image: "Figure A shows the location of the lungs and airways in the body. The inset image shows a detailed cross-section of the bronchioles and alveoli. Figure B shows lungs damaged by COPD. The inset image shows a detailed cross-section of the damaged bronchioles and alveolar walls." by National Heart Lung and Blood Institute. License: [Public Domain](#)

## Pathophysiology of Emphysema

The part of the acinus that is affected by destruction or permanent dilation decides the subtype of emphysema, which includes:

### 1. Centrilobular (proximal acinar) emphysema

Of this type, there is abnormal destruction or dilation of the respiratory bronchiole.

- Common among **chronic smokers** and coal workers' pneumoconiosis
- The excess amount of neutrophils, due to cigarette smoking, results in the release of huge amounts of **elastase** into the lung parenchyma, which leads to the inability of alpha-1-antitrypsin (AAT) to maintain the integrity of the [tissue](#).
- There is a loss of respiratory bronchioles, mainly in the upper and middle lobes.

### 2. Panacinar emphysema

This type is defined as enlargement or destruction of all parts of the acinus.

- The mutation of the normal alpha-1-antitrypsin gene (Pi) to PiZZ is one of the most common homozygous genetic mutations seen in Caucasians.
- It can also be seen in centrilobular emphysema in smokers.

### 3. Paraseptal (distal acinar) emphysema

This type results in the destruction of the alveolar ducts. It usually results in spontaneous pneumothorax when patients are affected with this type alone. However, it may occur in combination with other types of emphysema.

## Epidemiology of Emphysema

**United States:** 18 cases per 1000 persons suffer from emphysema and 34 cases per 1000 persons suffer from chronic bronchitis.

**International:** The prevalence of [COPD](#) stage II or higher is **10.1%**.

**Mortality:** There are 64.3 deaths from COPD per 100,000 population in adults older than 25 years in the United States.

**Gender:** It used to be more prevalent among men because of smoking. However, the difference in prevalence between the two genders is decreasing.

## Presentation of Emphysema

### Symptoms of emphysema

Patients usually ignore their symptoms and adapt themselves to them as they start gradually and progress over the years. These symptoms include:

- A productive cough worse in the morning with clear-to-white sputum
- Acute chest illness
- [Dyspnea](#)
- [Wheezing](#)

### Signs of emphysema

Observed signs may be different depending on the severity and the stage of the disease. They include:

- Limitation of expiratory flow with relative preservation of inspiratory flow
- Tachypnea
- [Dyspnea](#) with mild exertion
- Increased respiratory rate
- Use of accessory respiratory muscles
- Paradoxical contraction of lower intercostal spaces



[Image:](#) "A lateral CXR of a person with emphysema. Note the barrel chest and flat diaphragm." by James Heilman, MD. License: [CC BY-SA 3.0](#)

- Cyanosis
- Elevated Jugular venous pressure

- Atrophy of limb musculature
- Peripheral edema
- Barrel chest
- Diffuse or focal [wheezing](#)
- Diminished breath sounds
- Hyper-resonance upon percussion
- Prolonged expiration
- Hyperinflation in chest radiograph

## Causes of Emphysema

Different risk factors may increase the risk of developing emphysema, these causes and risk factors include:

- Cigarette smoking
- [Alpha-1-Antitrypsin \(AAT\) deficiency syndromes](#)
- Intravenous drug users
- Immune deficiency syndromes
- Vasculitis
- Connective tissue disorders
- Salla disease

## Differential Diagnosis of Emphysema

**Congestive heart failure:** this disorder might produce wheezing and can be difficult to distinguish from emphysema. However, a history of orthopnea and paroxysmal dyspnea that is nocturnal with the existence of fine basal crackles, and typical findings on upper body radiographs, can lead to the diagnosis of congestive heart failure.

**Bronchiectasis:** patients with bronchiectasis have chronic production of copious sputum that is purulent, coarse crackles, and clubbing upon physical examination. Abnormal findings on upper body radiographs and CT scans will be observed.

**Bronchiolitis obliterans:** it is observed in younger persons who do not smoke and in persons with collagen-vascular diseases. A CT scan characteristically shows regions of mosaic attenuation without proof of generalized emphysema.

**Chronic asthma:** the delayed onset of severe asthma may be hard to distinguish in older patients, but the main difference is the **response to bronchodilators** and **normal diffusion on pulmonary function tests**.

## Diagnosis of Emphysema

### Laboratory studies

- **Arterial blood gas analysis:** COPD patients have **mild-to-moderate hypoxemia** with **no associated hypercapnia**. However, hypercapnia may develop, and the hypoxemia worsens as the disease progresses.
- **Hematocrit:** polycythemia may result from chronic hypoxemia. A hematocrit result in more than **47% of female** and more than **52% of males** indicates polycythemia.

- **Serum bicarbonate:** serum bicarbonate levels are useful in the follow-up of the disease progression.
- **Serum alpha-1-antitrypsin:** when the serum levels fall below the protective threshold value, which is **3 to 7 mmol/L**, the diagnosis of alpha-1-antitrypsin deficiency is confirmed.
- **Sputum evaluation:** the sputum is mucoid with macrophages as the predominant cells. The sputum becomes purulent with exacerbation and includes **excessive neutrophils** and a mixture of organisms visualized through Gram staining.

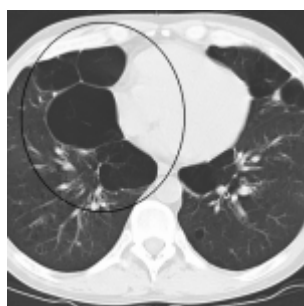
## Imaging studies



[Image:](#) "Patient with Severe Emphysema." by James Heilman, MD. License: [CC BY-SA 3.0](#)

### Chest radiograph:

Signs of **hyperinflation**, such as **diaphragm flattening**, **increased retrosternal air space**, and a **long narrow heart shadow** will be revealed on frontal and lateral chest x-rays. Other signs of emphysema include the **hyperlucency** of the lungs and **rapid tapering vascular shadows**. If the disease was complicated with pulmonary hypertension, the hilar vascular shadows become obvious and prominent. You may also notice **opacity** in the lower retrosternal air space and **right ventricular enlargement**.



[Image:](#) "Bullus emphysema as seen on CT" by James Heilman,

### CT scan:

High-resolution CT scans are also used in the diagnosis of emphysema as they are more **sensitive** than chest x-rays. It is highly **specific**, and it makes bullae visible and clear, which are usually hard to see on chest x-rays. CT scans are not always done for emphysema patients; however, they are **indicated for patients considered for surgical intervention**.

## Staging of Emphysema

**COPD** is divided into different stages depending on FEV<sub>1</sub>. The widely used staging system is the Global Initiative for Chronic Obstructive Lung Disease staging system, which is

- **Stage I (mild):** FEV<sub>1</sub> more than or equal to **80%** of the predicted value.
- **Stage II (moderate):** FEV<sub>1</sub> more than or equal to **50%** and less than **80%** of the predicted value.
- **Stage III (severe):** FEV<sub>1</sub> more than or equal to **30%** and less than **50%** of the predicted value.
- **Stage IV (very severe):** FEV<sub>1</sub> less than **50%** of the predicted value with chronic respiratory failure; or FEV<sub>1</sub> less than **30%** of the predicted value.

**Definition of respiratory failure:** PaO<sub>2</sub> less than 60 mm Hg (kPa 8.0) or a PaCO<sub>2</sub> higher than 50 mm Hg (kPa 6.7).

## Management of Emphysema

- **Smoking cessation:** it is the **most important factor** in the management of COPD patients.
- **Bronchodilators:** both short and long-acting bronchodilators can be used in the management of COPD, and they work by dilating airways, resulting in decreased airflow resistance.
- **Phosphodiesterase inhibitors:** they work by increasing intracellular cAMP, which results in **bronchodilation**.
- **Anti-inflammatory therapy:** inflammation is one of the important factors in the pathogenesis of COPD, therefore **oral and inhaled corticosteroids** are used to manage this inflammation.
- **Antibiotics:** chronic infection and colonization of the lower airways by bacteria may occur in COPD patients, which require the use of antibiotics.
- **Mucolytic agents:** they improve secretion clearance and reduce sputum viscosity.
- **Proton pump inhibitors:** studies showed that proton pump inhibitors reduce the incidence of the common cold in patients with COPD.
- **Oxygen therapy:** chronic hypoxemia may develop in COPD patients, which requires the use of oxygen therapy.

## Surgical intervention

- **Bullectomy:** this has been the **standard procedure** in selected patients for many years; it involves the removal of giant bullae.
- **Lung volume reduction surgery:** in this procedure, the most diseased parts of the lung are surgically resected to **decrease hyperinflation**.

- **Endobronchial valve placement:** it is still under investigation as a replacement for lung volume reduction surgery.
- **Lung transplantation:** most people who undergo lung transplantation are COPD patients.

## References

Carlo Raj. [Obstructive Lung Disease: Emphysema](#). Lecturio. Accessed March 7, 2016.

Kamran Boka. [Emphysema](#). Medscape. Updated October 23, 2014. Accessed 7, 2016.

[Emphysema](#). WebMD. Updated September 09, 2014. Accessed March 7, 2016.

Aquiles A Camelier, Daniel Hugo Winter, José Roberto Jardim, et al. Alpha-1 antitrypsin deficiency: diagnosis and treatment. SciELO.

**Legal Note:** Unless otherwise stated, all rights reserved by Lecturio GmbH. For further legal regulations see our [legal information page](#).

Notes