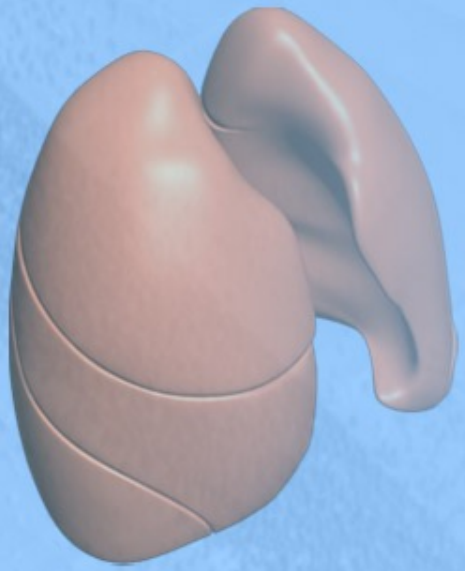
Pulmonary Disorders 2
Restrictive or Obstructive Lung Diseases

Dr. Gary Mumaugh

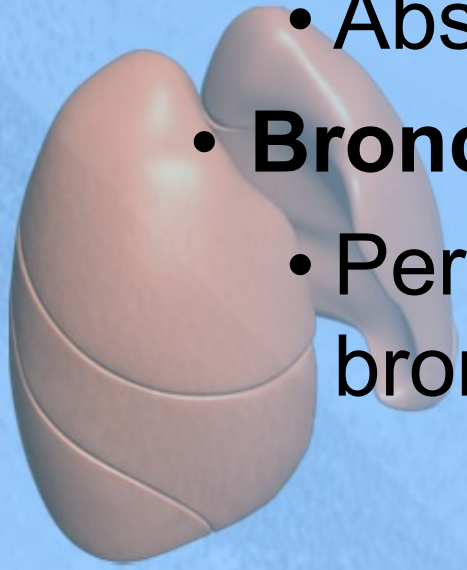


Major Pulmonary Topics

- Pleural Lesions
- Restrictive or Obstructive Lung Diseases
- Interstitial Lung Diseases
- Pulmonary Diseases of Vascular Origen
- Infections

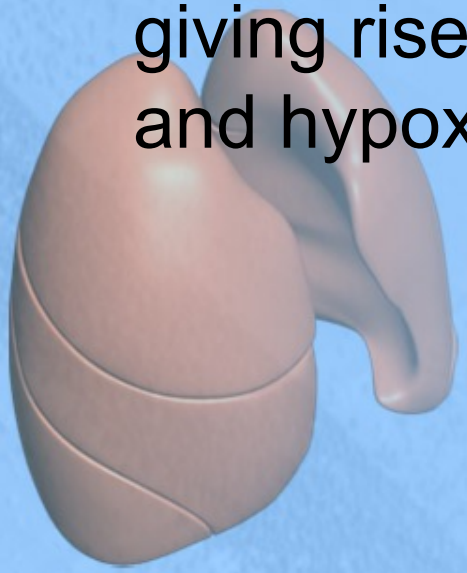
Restrictive Lung Diseases

- **Aspiration**
  - Passage of fluid and solid particles into the lungs
- **Atelectasis** - more detail next
  - Compression atelectasis
  - Absorption atelectasis
- **Bronchiectasis**
  - Persistent abnormal dilation of the bronchi



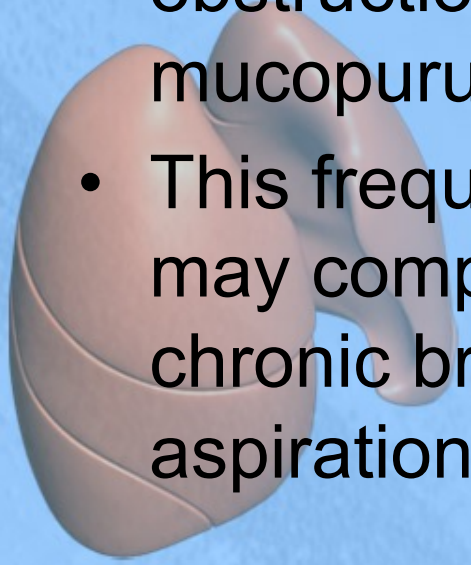
Atelectasis

- Atelectasis, also known as collapse, is loss of lung volume caused by inadequate expansion of air spaces.
- It results in shunting of inadequately oxygenated blood from pulmonary arteries into veins, thus giving rise to a ventilation- perfusion imbalance and hypoxia.



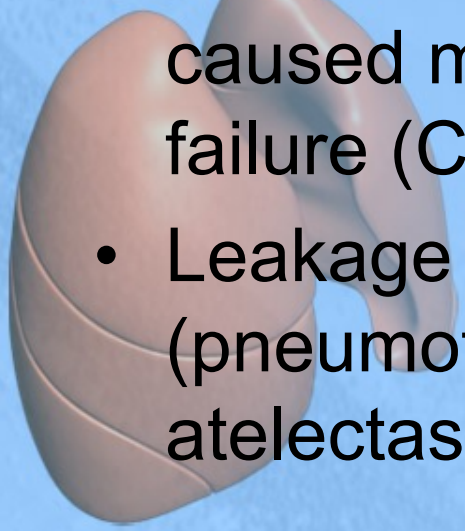
Resorption Atelectasis

- Resorption atelectasis occurs when an obstruction prevents air from reaching distal airways. The air already present gradually becomes absorbed, and alveolar collapse follows.
- The most common cause of resorption collapse is obstruction of a bronchus by a mucous or mucopurulent plug.
- This frequently occurs postoperatively but also may complicate bronchial asthma, bronchiectasis, chronic bronchitis, tumor, or foreign body aspiration, particularly in children.



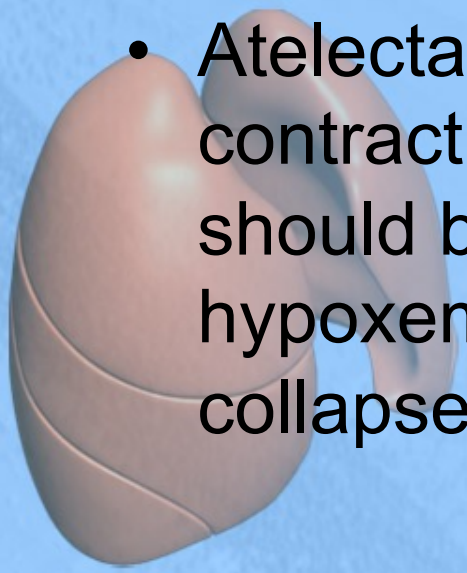
Compression Atelectasis

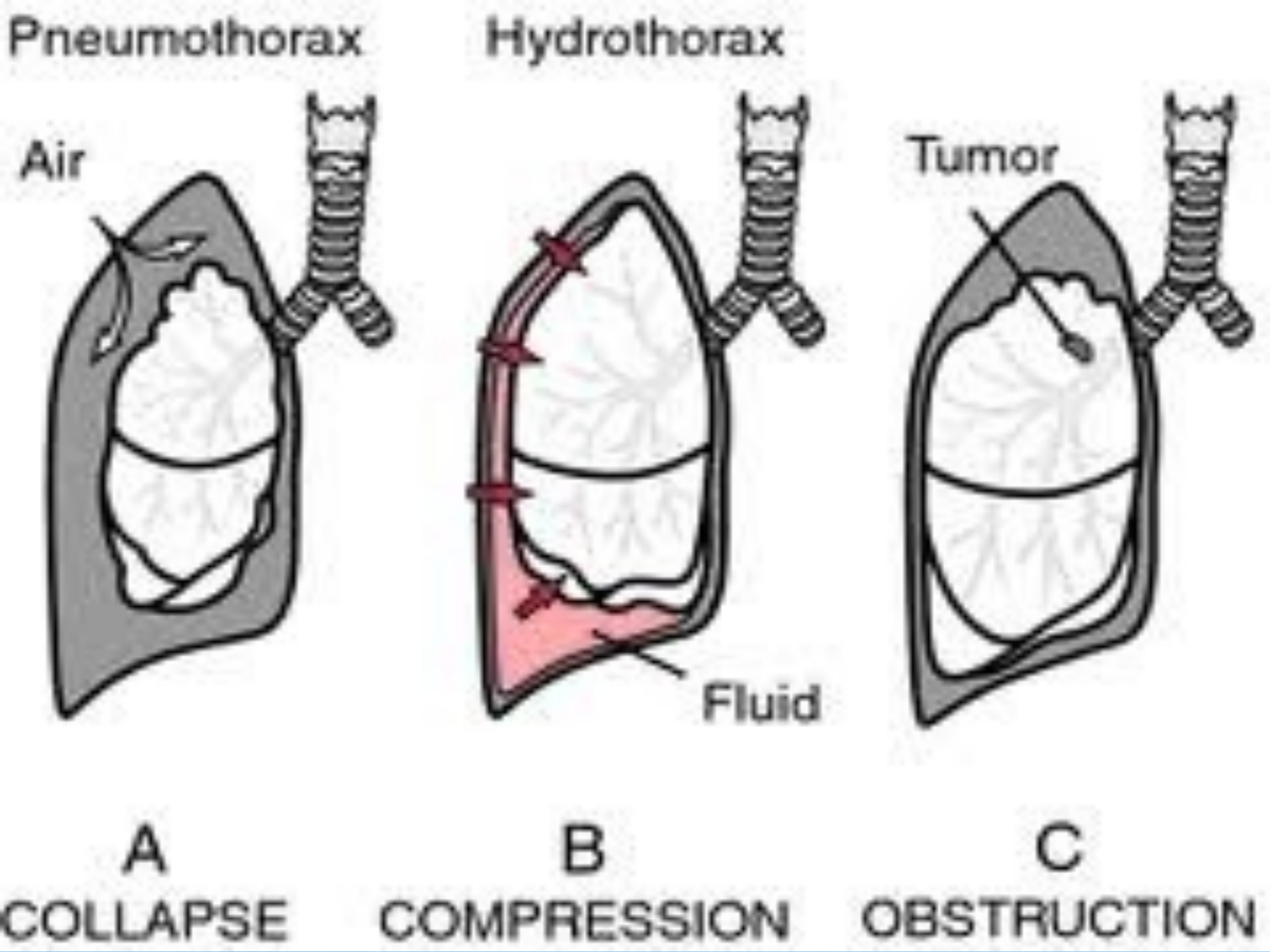
- Compression atelectasis is usually associated with accumulation of fluid, blood, or air within the pleural cavity, which mechanically collapses the adjacent lung.
- Sometimes called passive or relaxation atelectasis
- This is a frequent occurrence with pleural effusion, caused most commonly by congestive heart failure (CHF).
- Leakage of air into the pleural cavity (pneumothorax) also leads to compression atelectasis.



Contraction Atelectasis

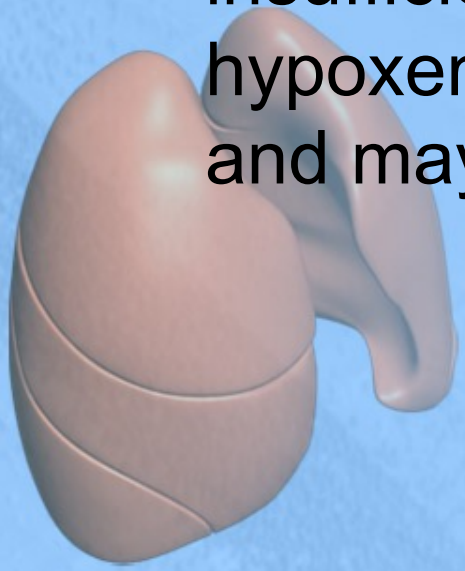
- Contraction occurs when either local or generalized fibrotic changes in the lung or pleura hamper expansion and increase elastic recoil during expiration.
- This is irreversible
- Atelectasis (except when caused by contraction) is potentially reversible and should be treated promptly to prevent hypoxemia and superimposed infection of the collapsed lung.



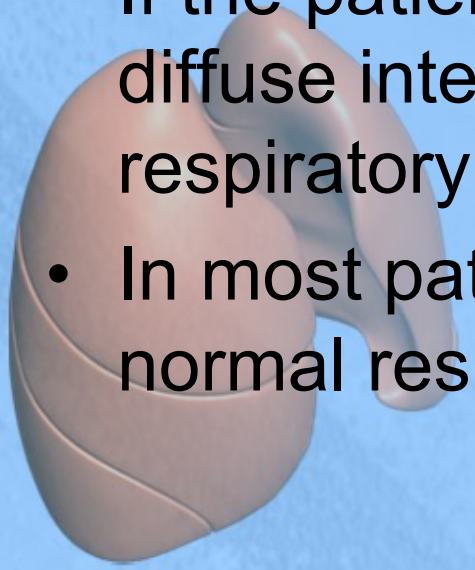

Pneumothorax
Air
Hydrothorax
Fluid
Tumor
A
COLLAPSE
B
COMPRESSION
C
OBSTRUCTION

Acute respiratory distress syndrome (ARDS)

- Acute respiratory distress syndrome (ARDS) is a clinical syndrome caused by diffuse alveolar capillary and epithelial damage.
- The usual course is characterized by rapid onset of life-threatening respiratory insufficiency, cyanosis, and severe arterial hypoxemia that is refractory to oxygen therapy and may progress to multisystem organ failure.

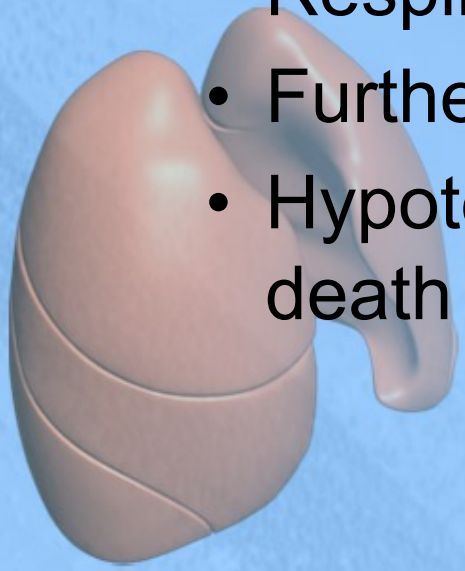


- About 85% of patients of patients with an acute lung injury will develop ARDS within 72 hours after the injury.
  - The mortality rate has decreased from 60% to 40% in the last decade.
  - Worse prognosis more common with age, bacteremia, sepsis, and multisystem failure.
- If the patient survives the initial acute phase, diffuse interstitial fibrosis can occur, compromising respiratory function.
- In most patients who survive the acute phase, normal respiratory function returns in 6-12 months.



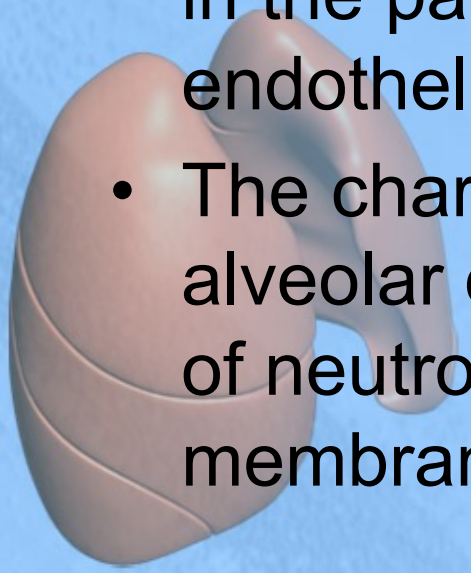
ARDS Manifestations

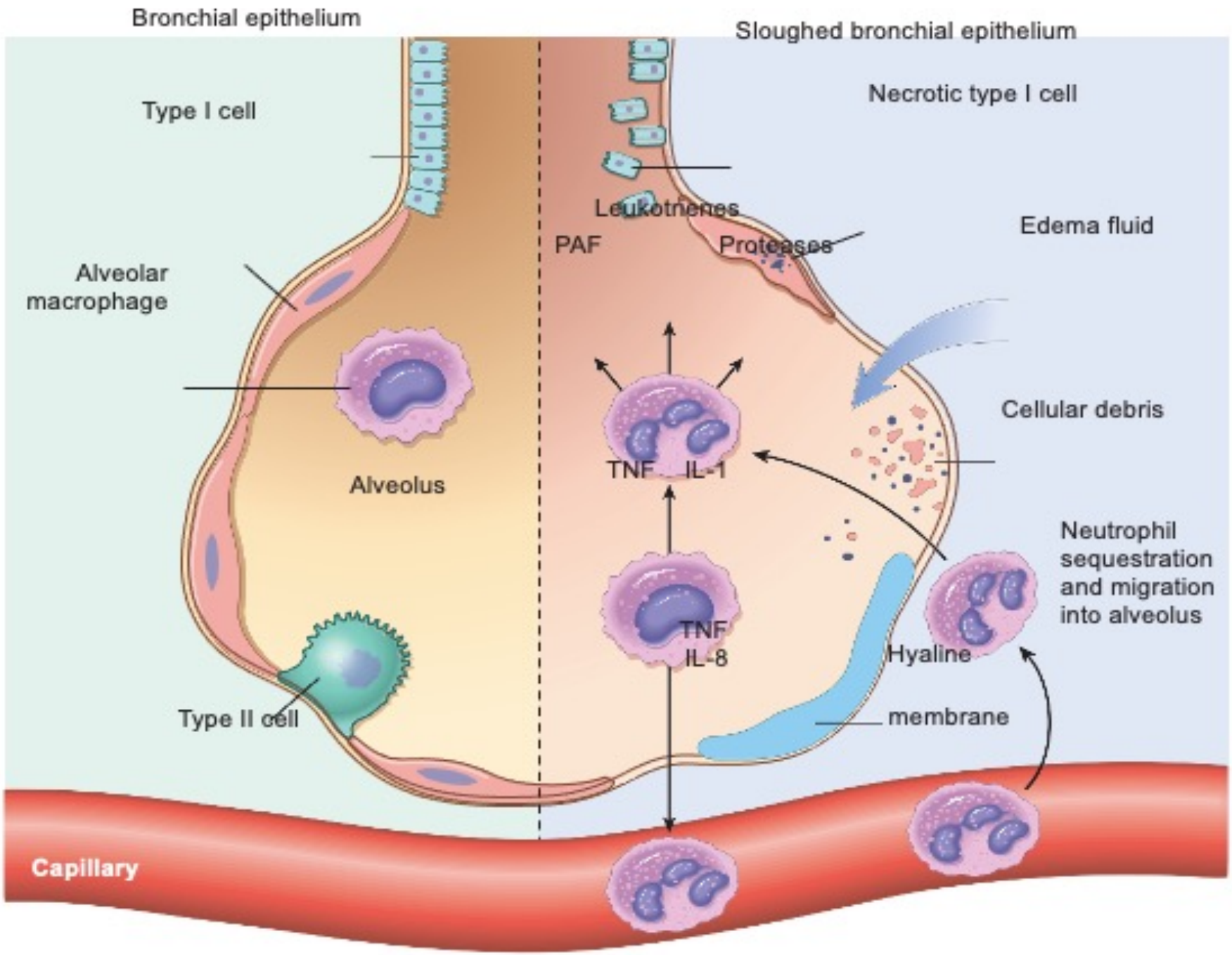
- Hyperventilation
- Respiratory alkalosis
- Dyspnea and hypoxemia
- Metabolic acidosis
- Hypoventilation
- Respiratory acidosis
- Further hypoxemia
- Hypotension, decreased cardiac output, death



ARDS Summary

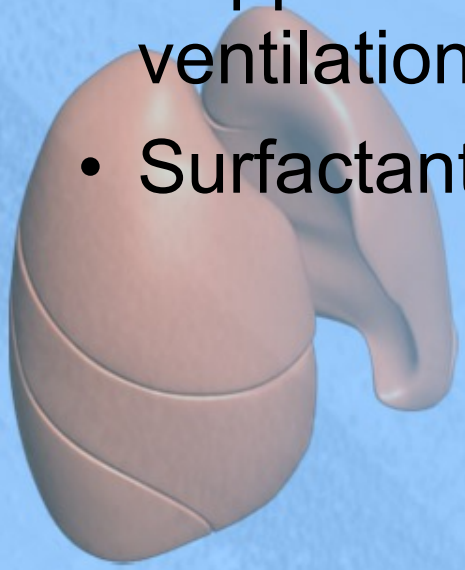
- ARDS is a clinical syndrome of progressive respiratory insufficiency caused by diffuse alveolar damage in the setting of sepsis, severe trauma, or diffuse pulmonary infection.
- Neutrophils and their products have a crucial role in the pathogenesis of ARDS by causing endothelial and epithelial injury.
- The characteristic histologic picture is that of alveolar edema, epithelial necrosis, accumulation of neutrophils, and presence of hyaline membranes lining the alveolar ducts.

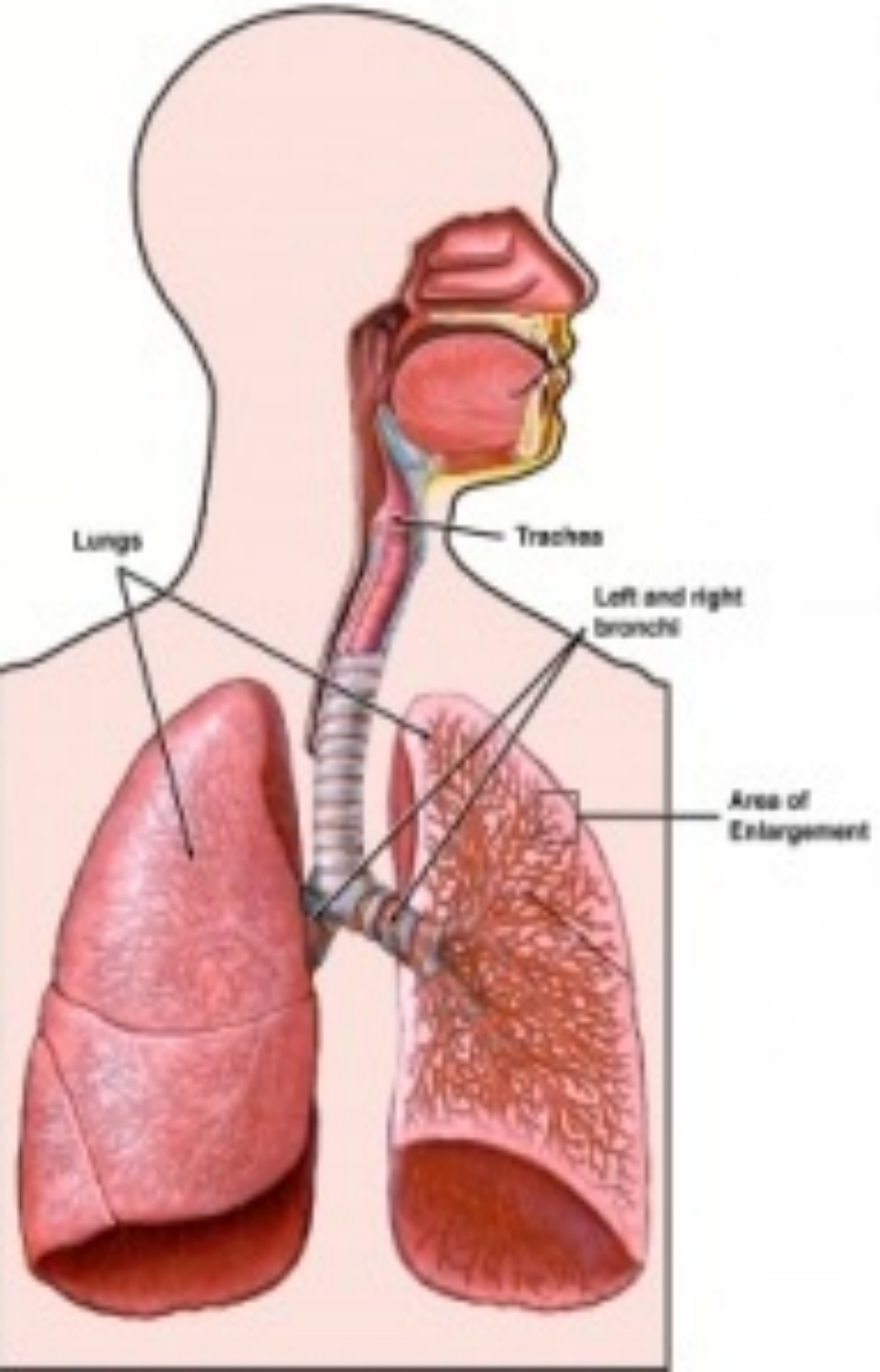



Bronchial epithelium
Sloughed bronchial epithelium
Type I cell
Necrotic type I cell
Leukotrienes
PAF
Proteases
Edema fluid
Alveolar macrophage
Alveolus
TNF
IL-1
Cellular debris
Neutrophil sequestration and migration into alveolus
TNF
IL-8
Hyaline membrane
Type II cell
Capillary

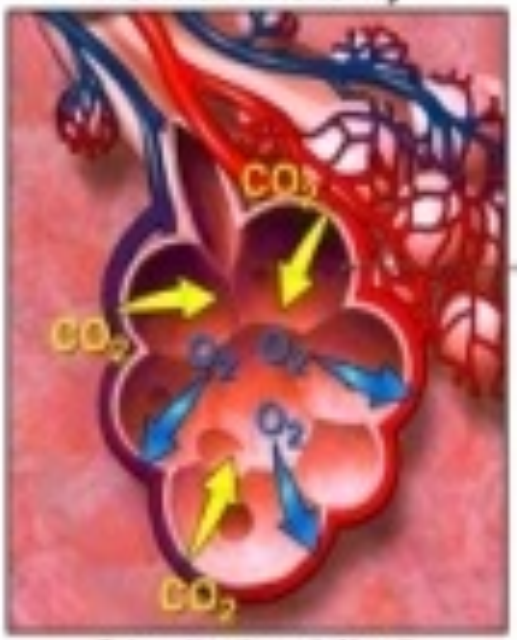
Acute respiratory distress syndrome (ARDS) Evaluation and treatment

- Physical examination, blood gases, and radiologic examination
- Supportive therapy with oxygenation and ventilation and prevention of infection
- Surfactant to improve compliance





Normal Anatomy



Normal gas exchange across thin alveolar walls allowing the uptake of fresh oxygen and the release of carbon dioxide

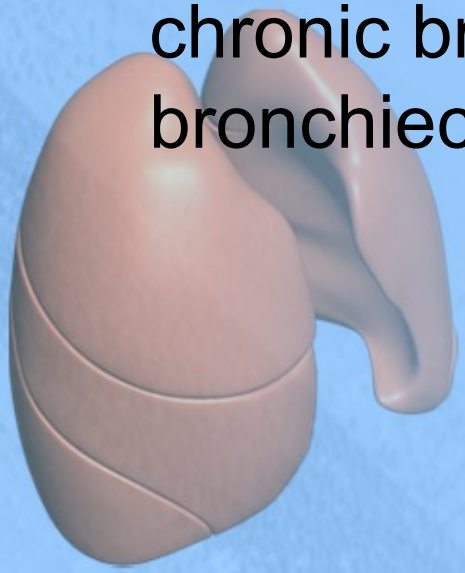
Cut-section through Alveoli at Terminus of Bronchi

ARDS

Fluid releasing from capillaries filling the alveolar space and preventing gas exchange

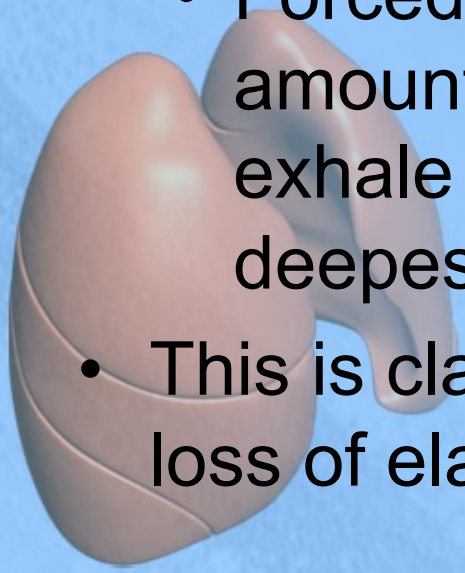
Obstructive Lung Diseases

- Obstructive airway disease is characterized by limitation of airflow, usually resulting from an increase in resistance by a partial or complete obstruction at any level.
- The major obstructive disorders are asthma, chronic bronchitis, emphysema, and bronchiectasis.



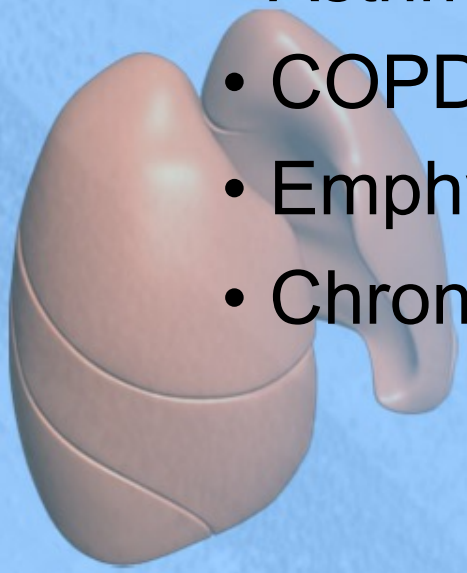
Obstructive Lung Diseases

- In patients with these diseases, forced vital capacity (FVC) is either normal or slightly decreased, while the expiratory flow rate, usually measured as the forced expiratory is significantly decreased.
  - Forced vital capacity or FVC, which is the amount of air that an individual can forcibly exhale from his / her lungs after taking the deepest breath they can.
- This is classically observed in asthma, or from loss of elastic recoil, characteristic of emphysema.



Pulmonary Disorders

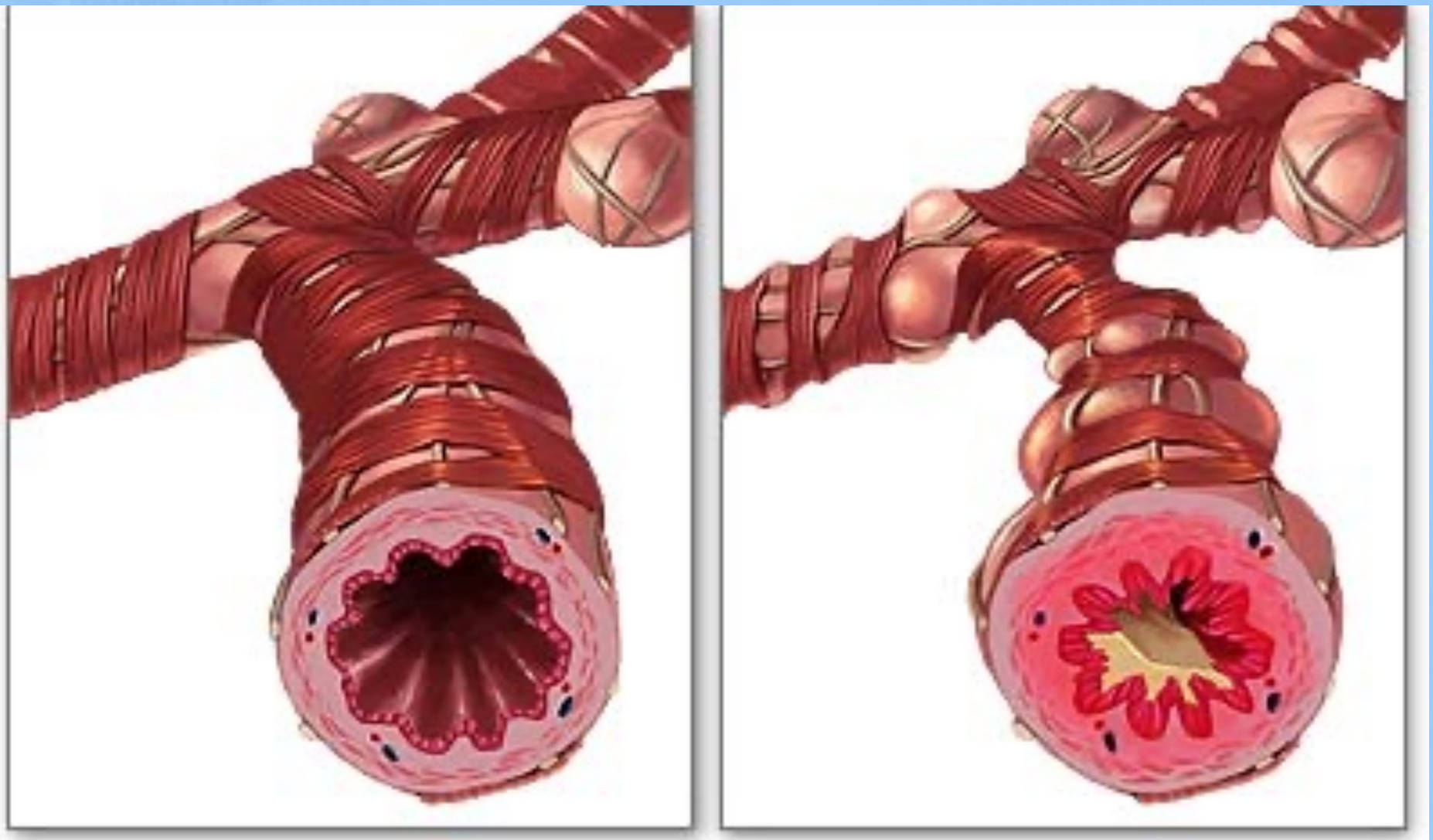
- **Obstructive lung diseases**
  - Airway obstruction that is worse with expiration
  - Common signs and symptoms
    - Dyspnea and wheezing
  - Common obstructive disorders:
    - Asthma
    - COPD
    - Emphysema
    - Chronic bronchitis



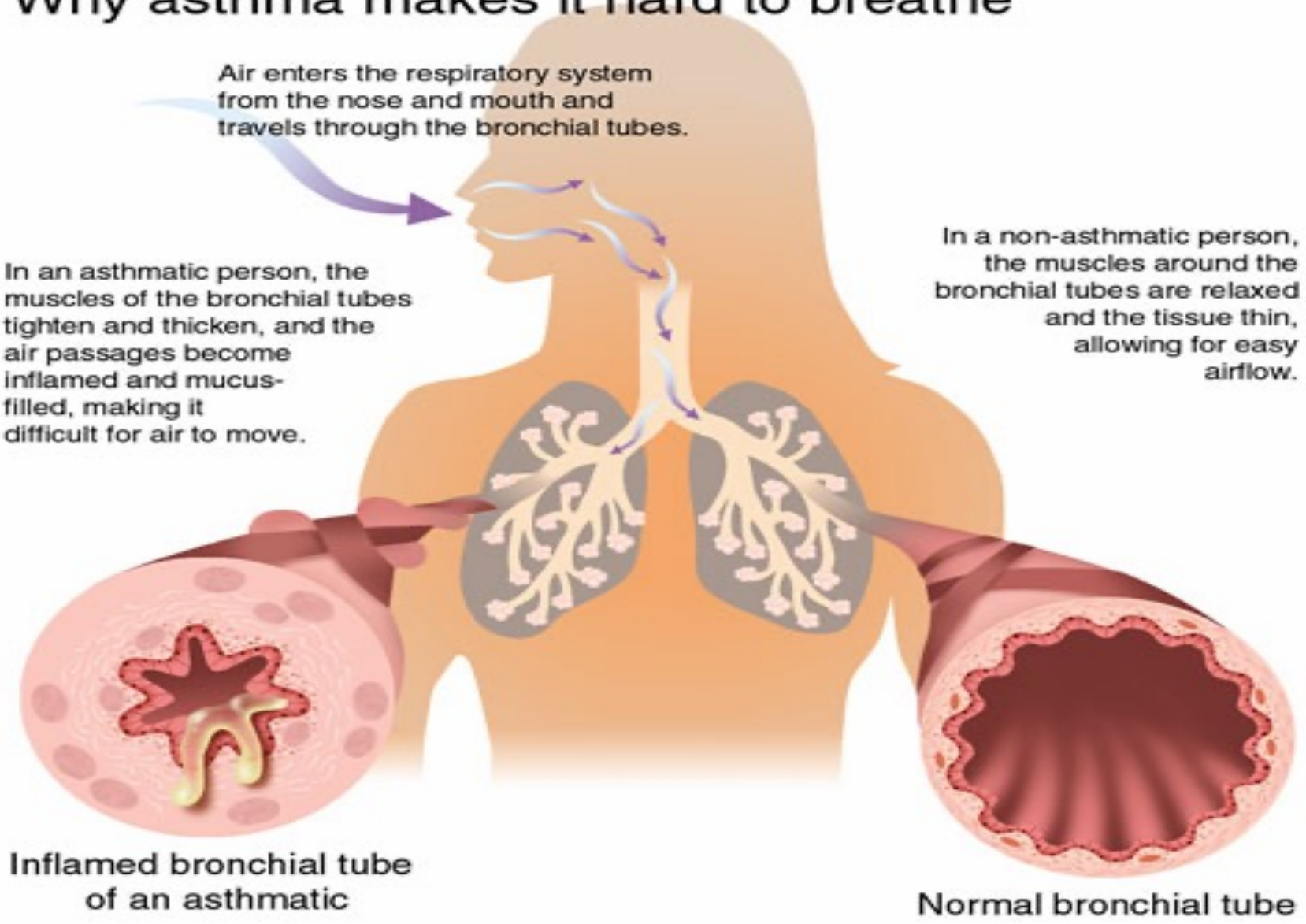
Disorders Associated with Airway Flow

Clinical Entity	Anatomic Site	Major Pathologic Changes	Etiology	Signs/Symptoms
Chronic bronchitis	Bronchus	Mucous gland hypertrophy and hyperplasia, hypersecretion	Tobacco smoke, air pollutants	Cough, sputum production
Bronchiectasis	Bronchus	Airway dilation and scarring	Persistent or severe infections	Cough, purulent sputum, fever
Asthma	Bronchus	Smooth muscle hypertrophy and hyperplasia, excessive mucus, inflammation	Immunologic or undefined causes	Episodic wheezing, cough, dyspnea
Emphysema	Acinus	Air space enlargement, wall destruction	Tobacco smoke	Dyspnea
Small airway disease, bronchiolitis*	Bronchiole	Inflammatory scarring, partial obliteration of bronchioles	Tobacco smoke, air pollutants	Cough, dyspnea

Asthma



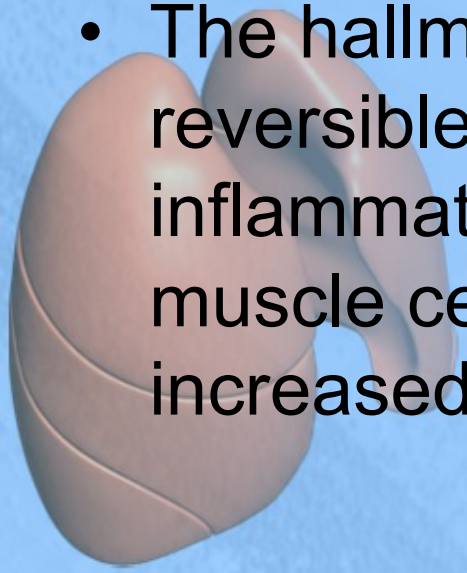
Why asthma makes it hard to breathe



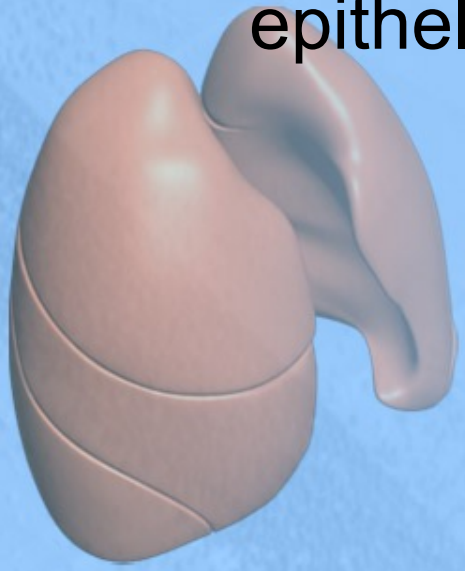
Source: American Academy of Allergy, Asthma and Immunology

Asthma

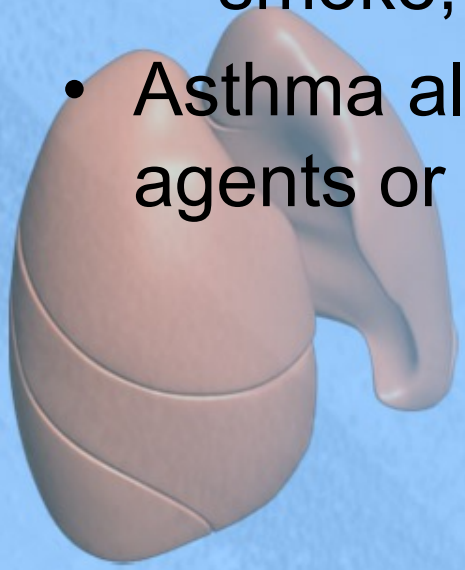
- Asthma is a chronic inflammatory disorder of the airways that causes recurrent episodes of wheezing, breathlessness, chest tightness, and cough, particularly at night and/or early in the morning.
- The hallmarks of the disease are intermittent and reversible airway obstruction, chronic bronchial inflammation with eosinophils, bronchial smooth muscle cell hypertrophy and hyperreactivity, and increased mucus secretion.



- Some of the stimuli that trigger attacks in patients would have little or no effect in persons with normal airways.
  - Many cells play a role in the inflammatory response, in particular eosinophils, mast cells, macrophages, lymphocytes, neutrophils, and epithelial cells.

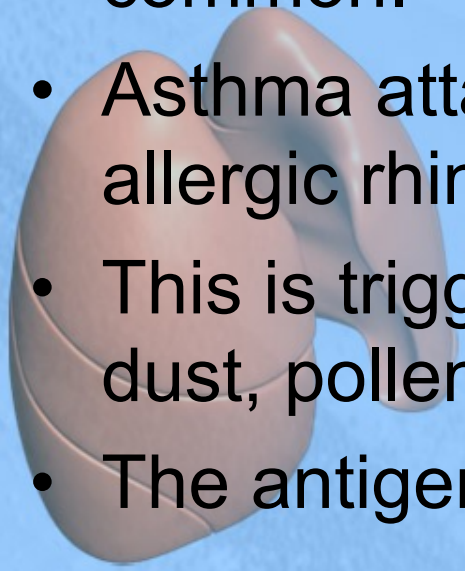


- Asthma may be categorized into *atopic* (evidence of allergen sensitization, often in a patient with a history of allergic rhinitis, eczema) and *nonatopic*.
  - In either type, episodes of bronchospasm can be triggered by diverse mechanisms, such as respiratory infections (especially viral), environmental exposure to irritants (e.g., smoke, fumes), cold air, stress, and exercise.
- Asthma also may be classified according to the agents or events that trigger bronchoconstriction.

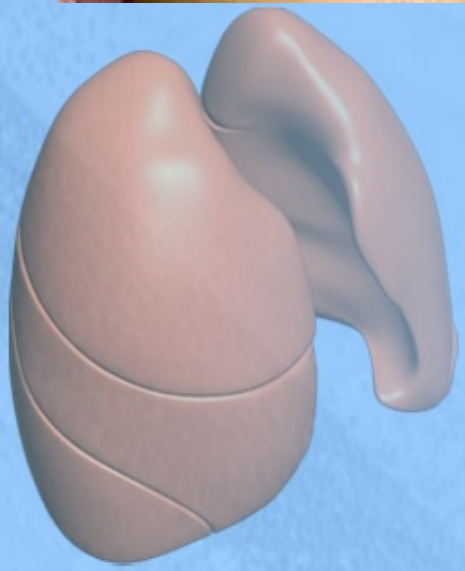


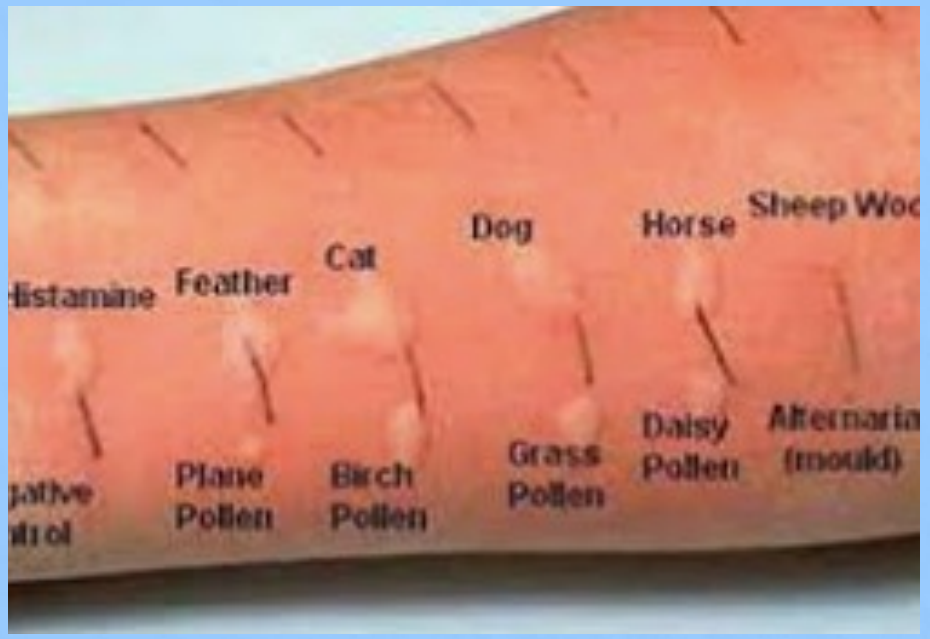
Atopic Astma

- This is the most common type of asthma, usually staring in childhood and is a class Type 1 Hypersensitivity reaction.
- This is sometimes called extrinsic asthma because it is an allergic reaction due to something outside
- A positive family history of atopy and or asthma is common.
- Asthma attacks in children are often preceded by allergic rhinitis, urticaria, or eczema.
- This is triggered by environmental antigens, such as dust, pollens animal dander and foods.
- The antigens are diagnosed with skin tests.



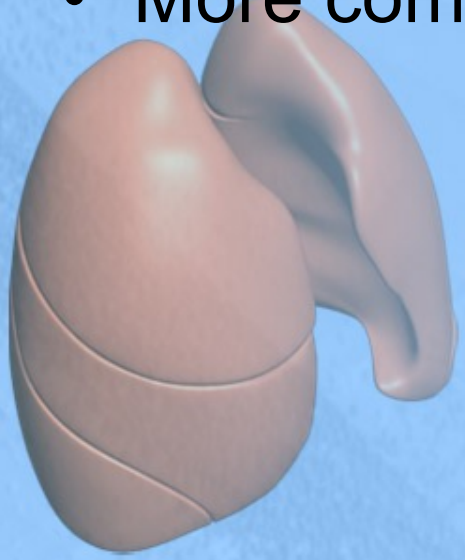

Positive Reaction



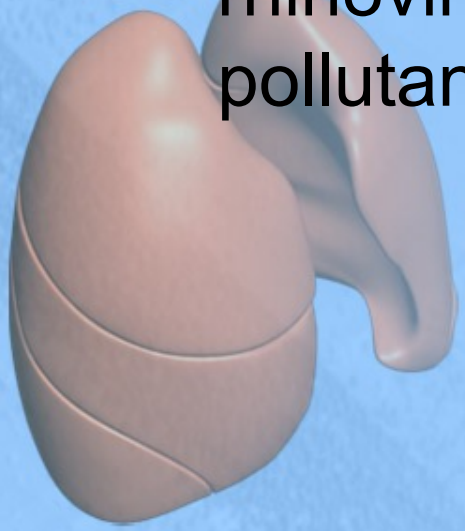

Feather
Cat
Dog
Horse
Plane Pollen
Birch Pollen
Grass Pollen
Daisy Pollen

Non Atopic Asthma

- Sometimes called intrinsic asthma because it is caused by something inside.
- Viral infection, irritants like epithelial damage, mucosal inflammation, emotional upset, parasympathetic input.
- More common in adults

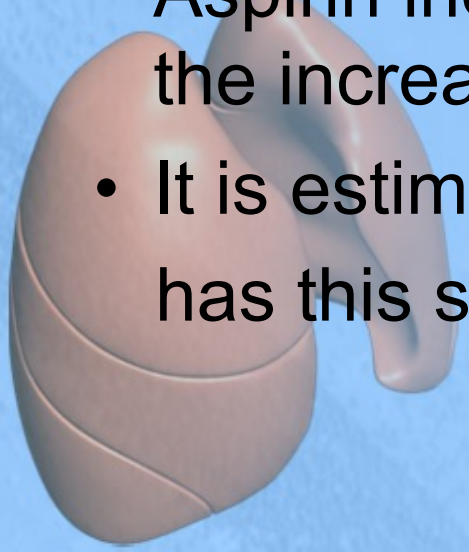


- Patients with nonatopic forms of asthma do not have evidence of allergen sensitization, and skin test results usually are negative. A positive family history of asthma is less common.
- Respiratory infections due to viruses (e.g., rhinovirus, influenza virus) and inhaled air pollutants are common triggers.

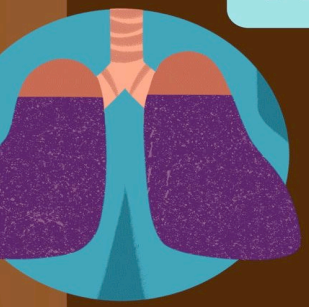


Drug Induced Asthma

- Several pharmacologic agents provoke asthma, aspirin being the most striking example.
- Patients with aspirin sensitivity present with recurrent rhinitis and nasal polyps, urticaria, and bronchospasm.
- Aspirin induces arachidonic acid metabolism and the increase of bronchoconstrictor leukotrienes.
- It is estimated that 10% of the adult population has this sensitivity.



Aspirin-Induced Asthma (AIA) Symptoms



hortness of breath

Wheezing

Hives

Facial flushing

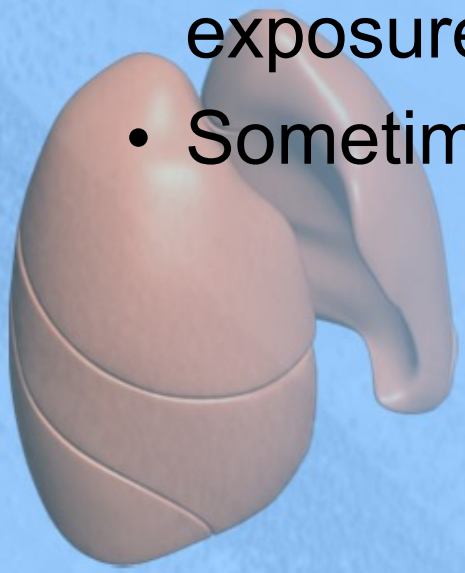
Coughing

Congestion

rywell

Occupational Asthma

- This form of asthma is stimulated by fumes (epoxy resins, plastics), organic and chemical dusts (wood, cotton, platinum), gases (toluene), and other chemicals.
- Asthma attacks usually develop after repeated exposure to the inciting antigen(s)
- Sometimes called wood dust asthma.



Indoor Air Pollutants

Corn starch powder carrier

Eliminated

Predictors of Positive Inhalation Challenges

rHev b5, rHev b 6.01, rHev b 6.02

Latex-specific IgE inflammation

Reduction in latex products

Occupational Asthma

Outdoor Air Pollutants

Nonallergic inflammation

Neutrophilic, Th1/Th17, AHR

Allergic inflammation:

Eosinophilic, AHR

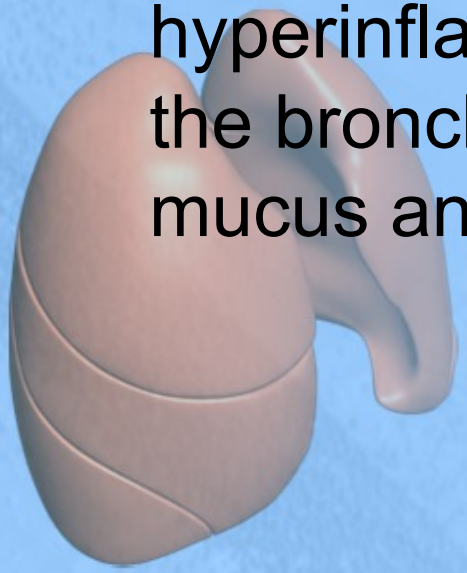
Early life

Complexity of Exposures

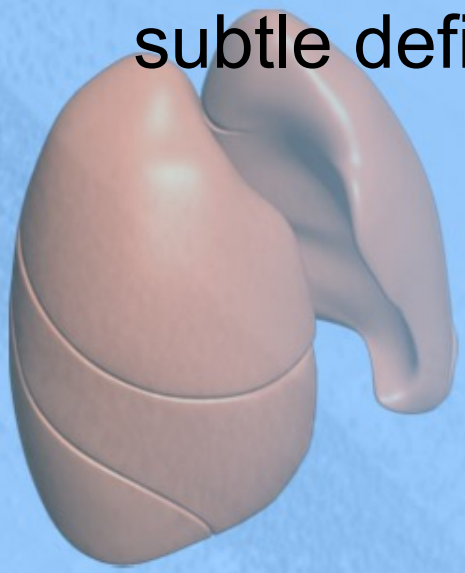
gram positive & gram negative bacteria, fungi, particulates, gases, pesticides, allergens

Asthma Clinical Features

- An attack of asthma is characterized by severe dyspnea with wheezing; the chief difficulty lies in expiration.
- The victim labors to get air into the lungs and then cannot get it out, so that there is progressive hyperinflation of the lungs with air trapped distal to the bronchi, which are constricted and filled with mucus and debris.

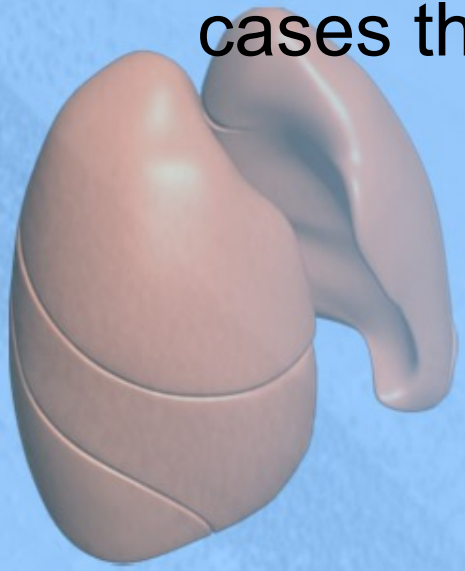


- In the usual case, attacks last from 1 to several hours and subside either spontaneously or with therapy, usually bronchodilators and corticosteroids.
- Intervals between attacks are characteristically free from overt respiratory difficulties, but persistent, subtle deficits can be detected by spirometry.



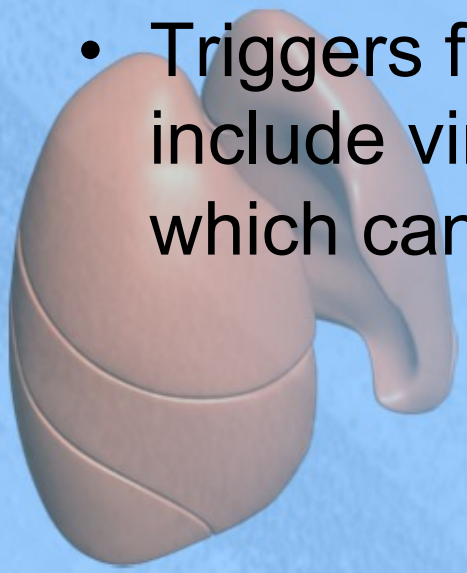
Status Astmaticus

- Occasionally a severe paroxysm occurs that does not respond to therapy and persists for days and even weeks (*status asthmaticus*).
- The associated hypercapnia, acidosis, and severe hypoxia may be fatal, although in most cases the condition is more disabling than lethal.

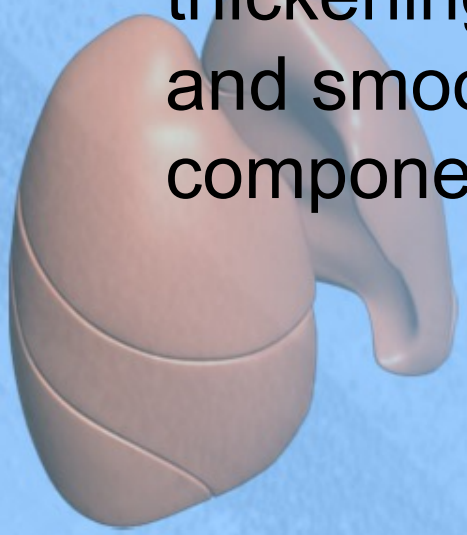


Asthma Summary

- Asthma is characterized by reversible bronchoconstriction caused by airway hyperresponsiveness to a variety of stimuli.
- Atopic asthma is caused by a IgE-mediated immunologic reaction to environmental allergens.
- Triggers for nonatopic asthma are less clear but include viral infections and inhaled air pollutants, which can also trigger atopic asthma.

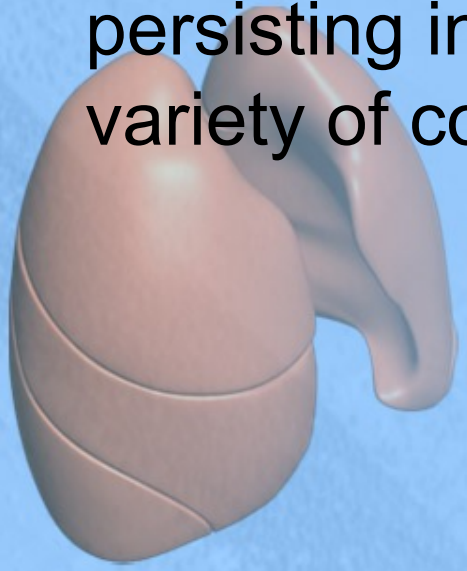


- Eosinophils are key inflammatory cells found in almost all subtypes of asthma; eosinophil products such as major basic protein are responsible for airway damage.
- Airway remodeling (sub-basement membrane thickening and hypertrophy of bronchial glands and smooth muscle) adds an irreversible component to the obstructive disease.



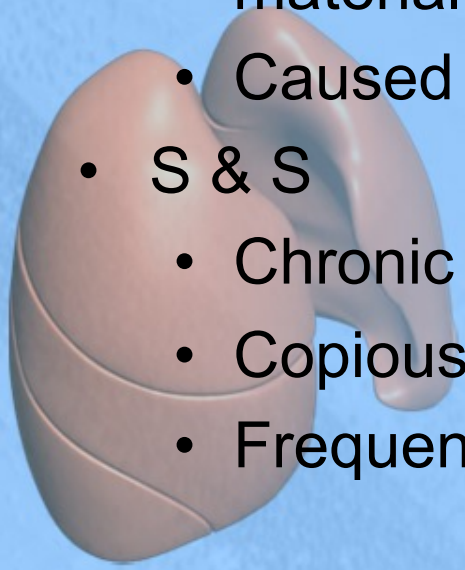
Bronchiectasis

- Bronchiectasis is the permanent dilation of bronchi and bronchioles caused by destruction of the muscle and the supporting elastic tissue, resulting from or associated with chronic necrotizing infections.
- It is not a primary disease but rather secondary to persisting infection or obstruction caused by a variety of conditions.

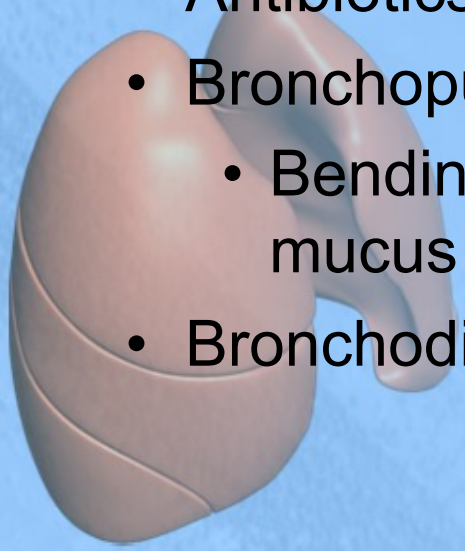


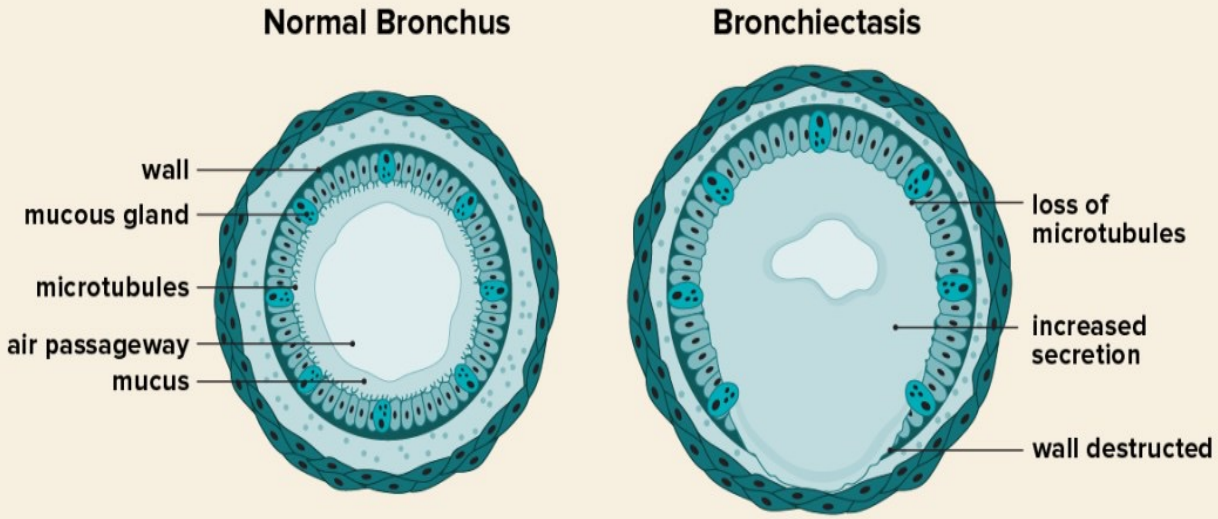
Bronchiectasis

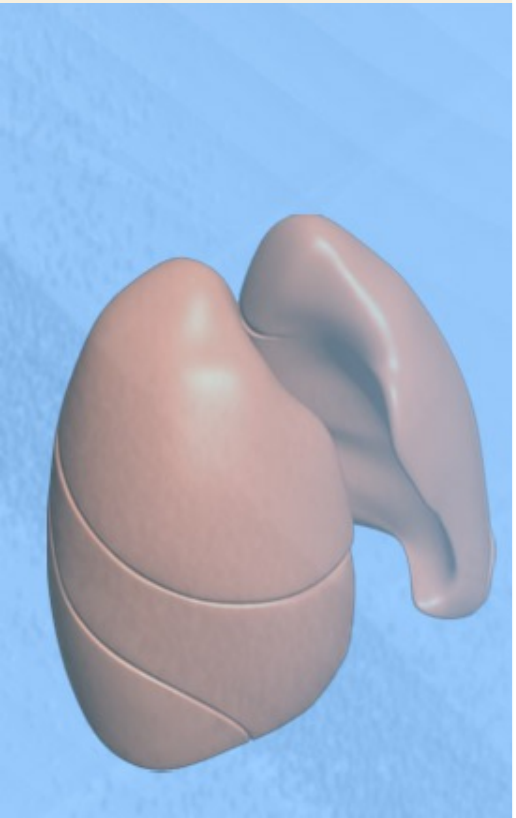
- Pathophysiology
  - Irreversible dilation of part of the bronchial tree
  - Caused by chronic infection of bronchi & bronchioles
  - Chronic bronchial infection causes a dilatation of the air passages which are trapped with muco-purulent material
  - Caused by slow-growing bacteria and fungi
- S & S
  - Chronic deep hacking cough
  - Copious amounts of foul-spelling pus sputum
  - Frequent attacks of pneumonia

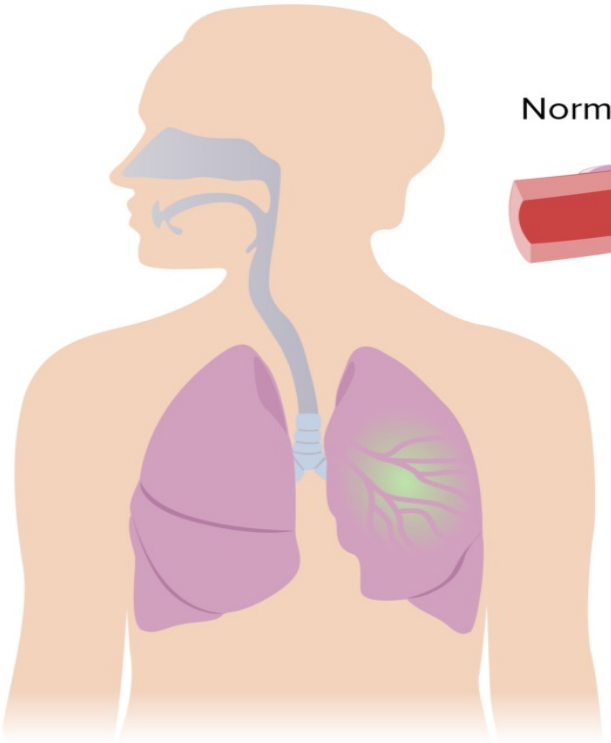


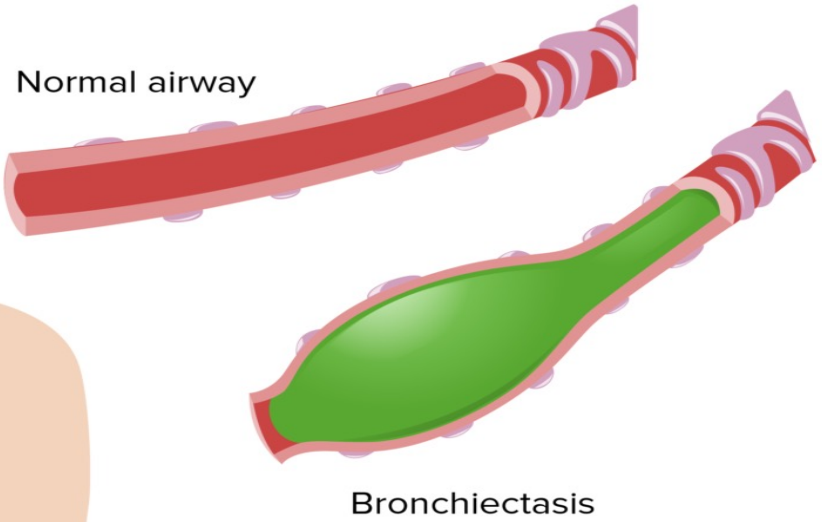
- Diagnosis
  - Localized rales and coarse ronchi
  - Appears similar to COPD with clubbing
  - Normal blood gases
  - History of chronic infection
  - CT scan confirms the diagnosis
- Treatment
  - Antibiotics – ciprofloxacin
  - Bronchopulmonary drainage
    - Bending over, almost standing on head, to get the mucus up and out
  - Bronchodilators



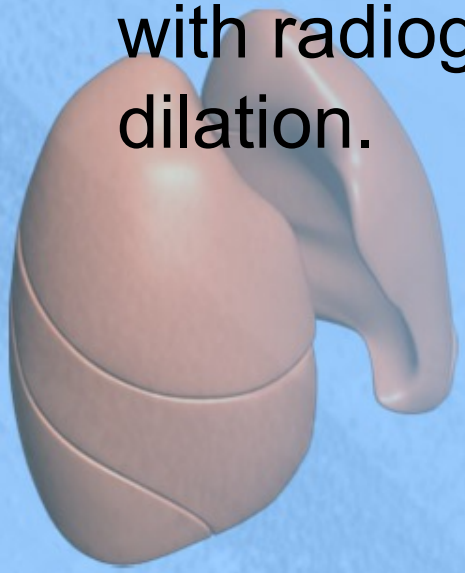

Normal Bronchus
Bronchiectasis
wall
mucous gland
microtubules
air passageway
mucus
loss of microtubules
increased secretion
wall destructed





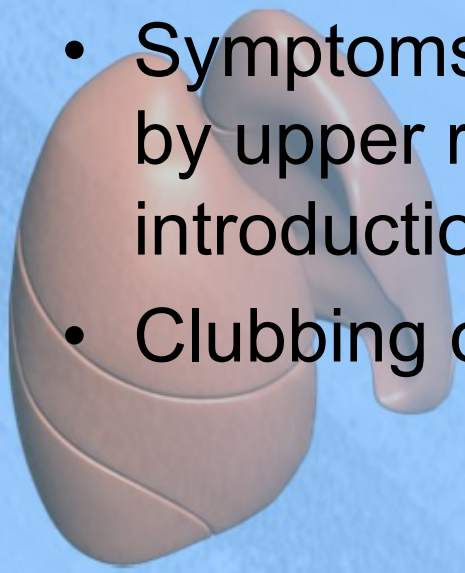

Normal airway
Bronchiectasis

- Once developed, it gives rise to a characteristic symptom complex dominated by cough and expectoration of copious amounts of purulent sputum.
- Diagnosis depends on an appropriate history along with radiographic demonstration of bronchial dilation.



Clinical Features of Bronchiectasis

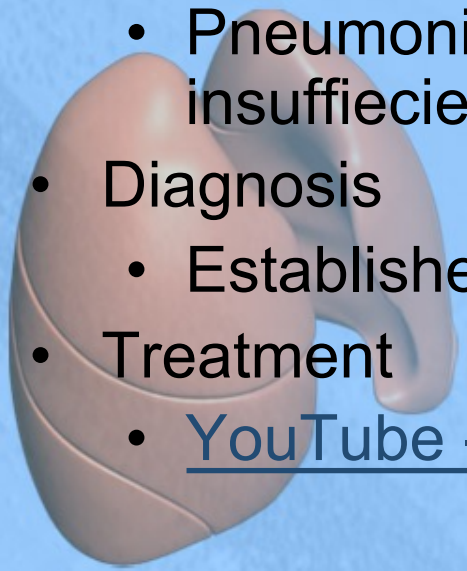
- The clinical manifestations consist of severe, persistent cough with expectoration of mucopurulent, sometimes fetid, sputum.
  - The sputum may contain flecks of blood; frank hemoptysis can occur.
- Symptoms often are episodic and are precipitated by upper respiratory tract infections or the introduction of new pathogenic agents.
- Clubbing of the fingers may develop.

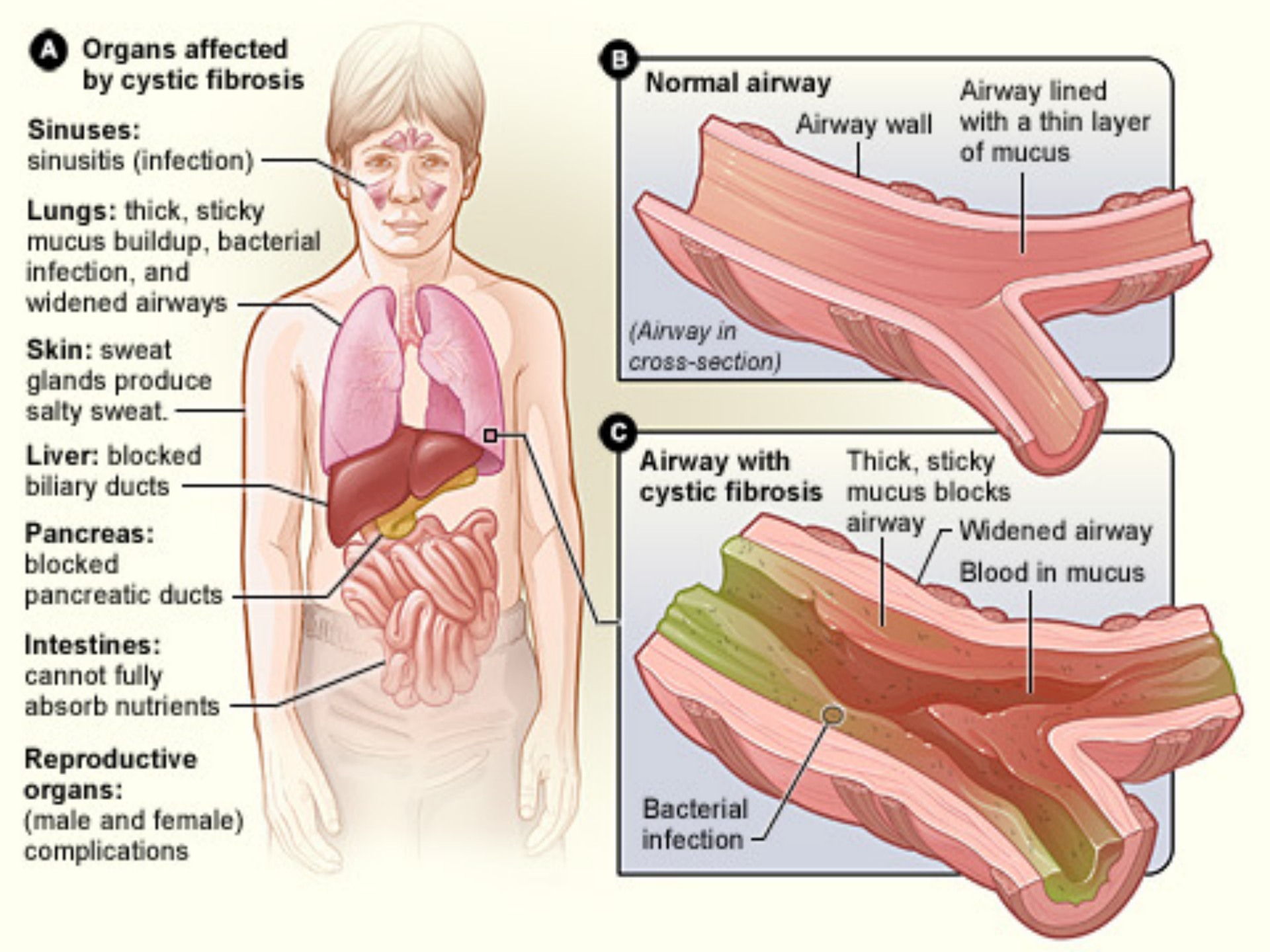


- The conditions that most commonly predispose to bronchiectasis include:
  - In *cystic fibrosis,* widespread severe bronchiectasis results from obstruction caused by the secretion of abnormally viscid mucus thus predisposing to infections of the bronchial tree.
  - In immunodeficiency states, particularly immunoglobulin deficiencies, localized or diffuse bronchiectasis is likely to develop because of an increased susceptibility to repeated bacterial infections.

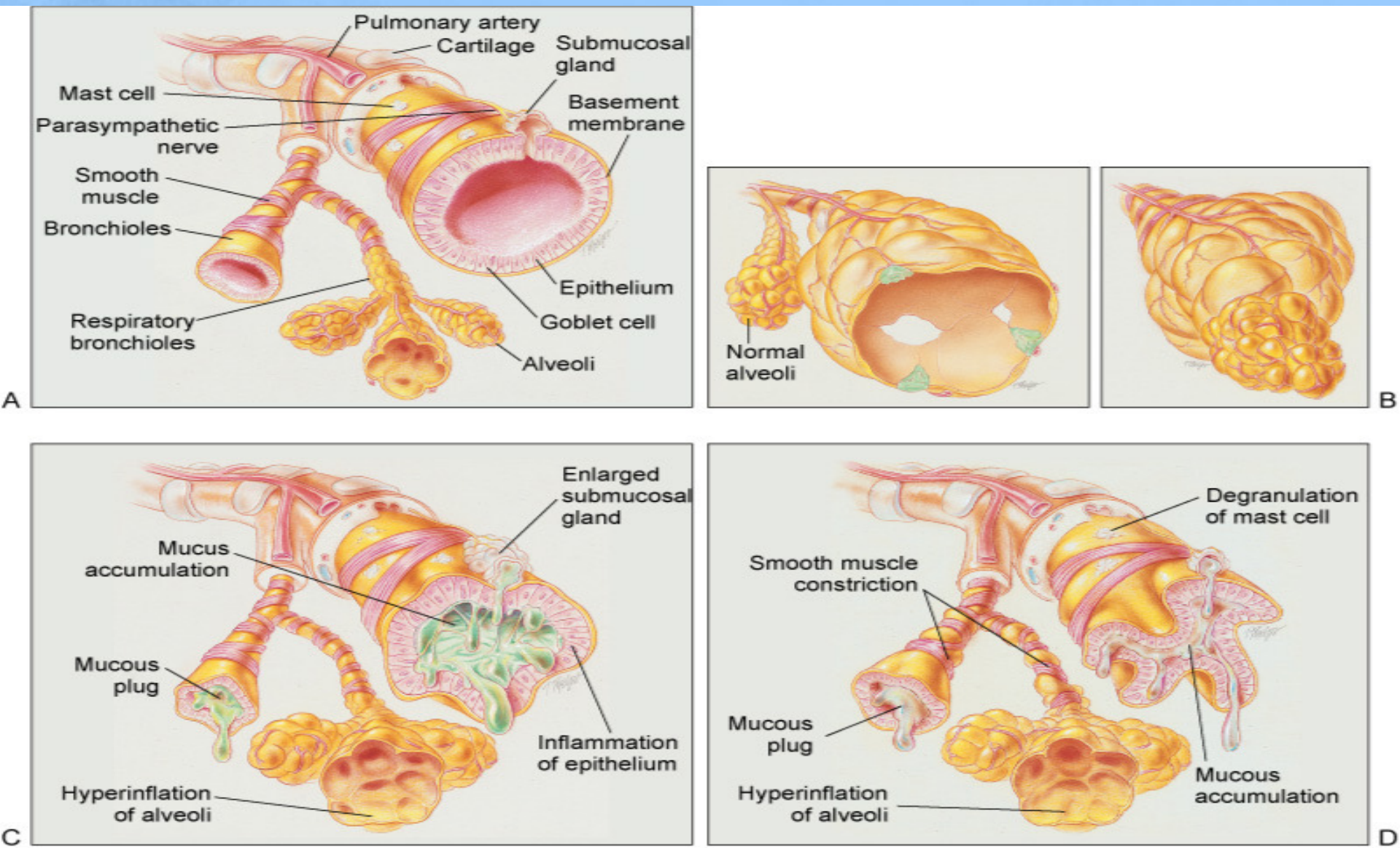
Cystic Fibrosis

- Inherited disease that causes thick, sticky mucus to build up in the lungs and digestive tract
- The most common type of chronic lung disease in children and young adults
  - 1 in every 3,300 – most children and teenagers
- May result in early death
- S & S
  - Pneumonitis, bronchiectasis, lung abscesses, pancreatic insuffieciency
- Diagnosis
  - Established by the sweat electrolyte test
- Treatment
  - YouTube - Current TV presents 'Dying Young'

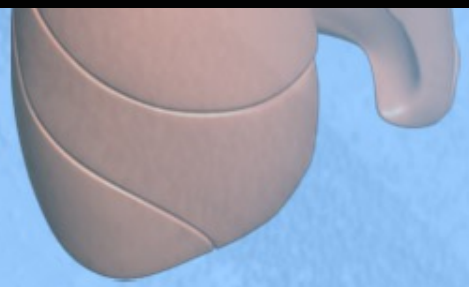



A Organs affected by cystic fibrosis
Sinuses: sinusitis (infection)
Lungs: thick, sticky mucus buildup, bacterial infection, and widened airways
Skin: sweat glands produce salty sweat.
Liver: blocked biliary ducts
Pancreas: blocked pancreatic ducts
Intestines: cannot fully absorb nutrients
Reproductive organs: (male and female) complications
B Normal airway
Airway wall
Airway lined with a thin layer of mucus
(Airway in cross-section)
C Airway with cystic fibrosis
Thick, sticky mucus blocks airway
Widened airway
Blood in mucus
Bacterial infection

Obstructive Pulmonary Disease

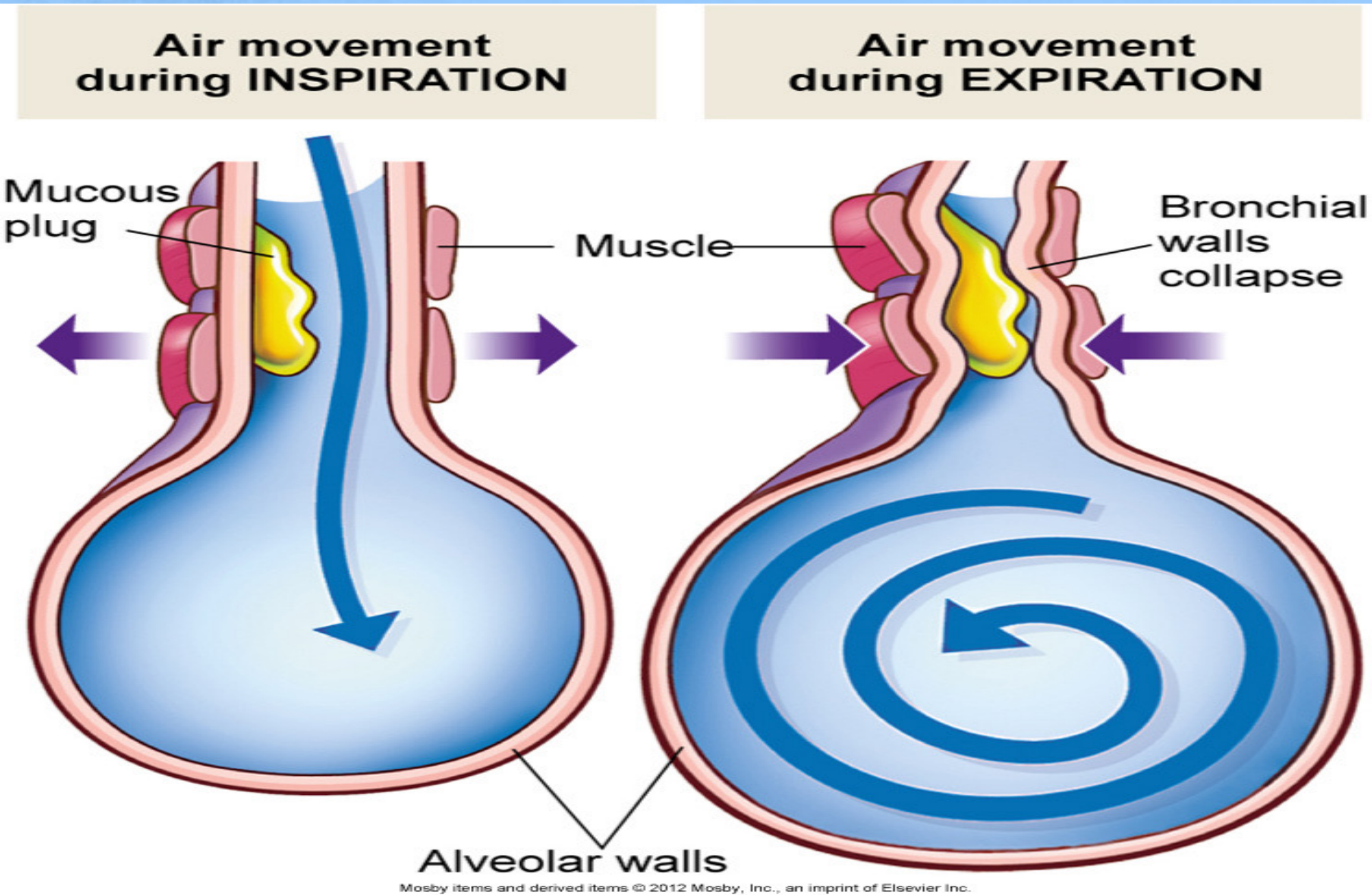


(Modified from Des Jardins T, Burton GG: *Clinical manifestations and assessment of respiratory disease*, ed 5, St Louis, 2006, Mosby.)



Struggle to Breath

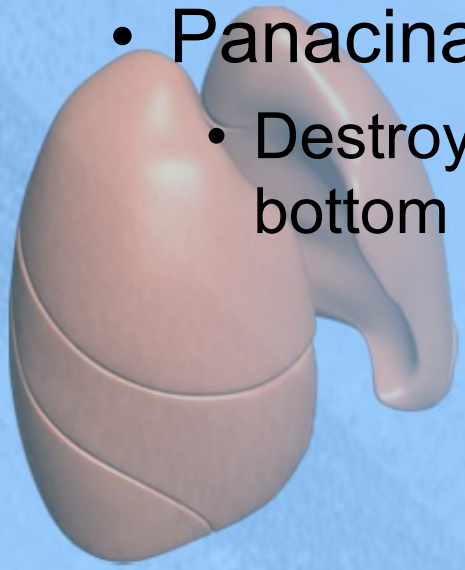
Chronic Obstructive Pulmonary Disease



Pulmonary Disorders

- **Obstructive lung diseases: chronic bronchitis**
  - Hypersecretion of mucus and chronic productive cough that lasts for at least 3 months of the year and for at least 2 consecutive years
  - Inspired irritants increase mucus production and the size and number of mucous glands
  - The mucus is thicker than normal
  - Bronchodilators, expectorants, and chest physical therapy used to treat

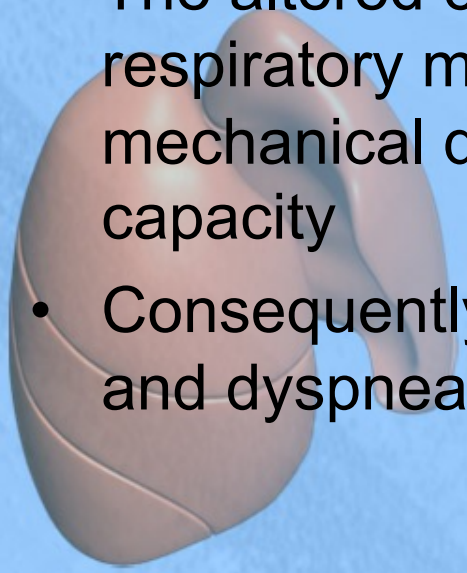
- **Obstructive lung diseases: emphysema**
  - Abnormal permanent enlargement of the gas-exchange airways accompanied by destruction of alveolar walls without obvious fibrosis
  - Loss of elastic recoil
  - Centriacinar emphysema
    - Most common form usually in proximal respiratory bronchioles in the top parts of the lungs
  - Panacinar emphysema
    - Destroys the entire alveolus and is usually found in the bottom half of the lungs

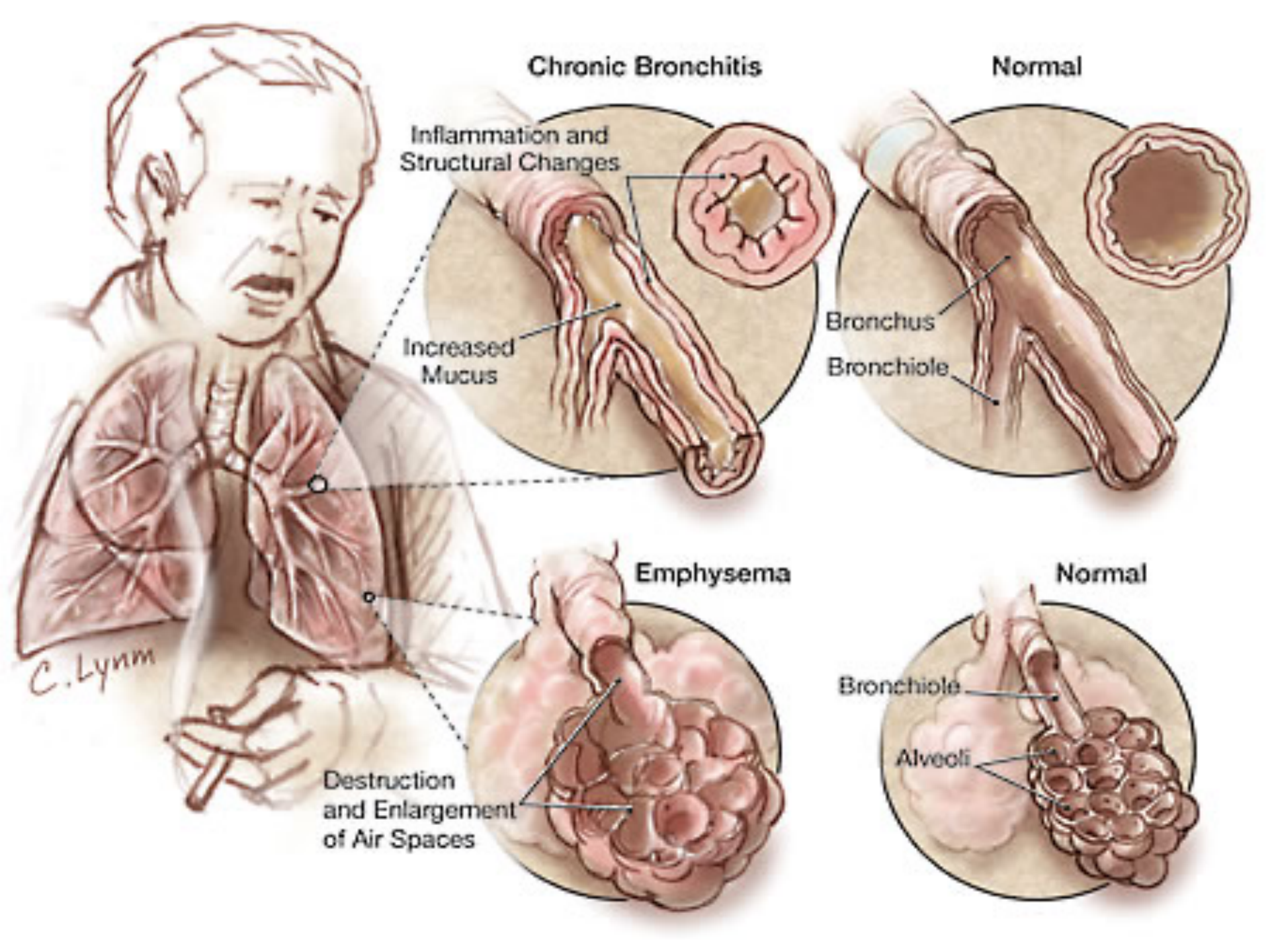


- How COPD develops
  - Smoking causes increased mucus production and bronchial inflammation
  - Nicotine paralyzes the mucociliary escalator
    - Mucociliary escalator traps mucus, bacteria, irritants
  - Nicotine blocks protein inhibitors which will eventually dissolve the alveoli
- Pathophysiology
  - Involves all four parts of the respiratory tract
    - Bronchi
    - Bronchioles
    - Alveoli
    - Parenchyma

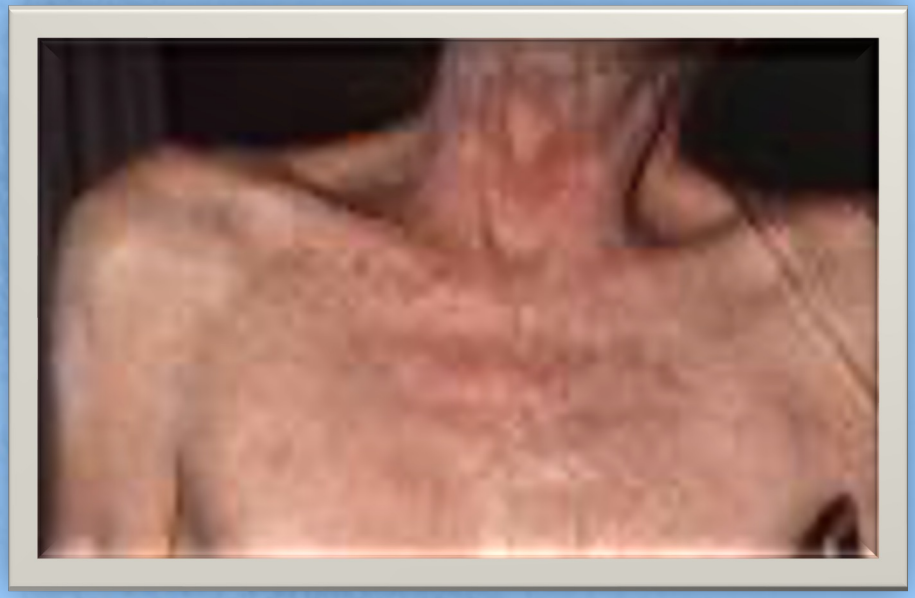
Specific Pathophysiology

- Increased resistance to airflow
- Loss of elastic recoil
- Decreased expiratory flow rate
- Alveolar walls frequently break because of the increased resistance of air flows
- The hyper inflated lungs flatten the curvature of the diaphragm and enlarge the rib cage
- The altered configuration of the chest cavity places the respiratory muscles, including the diaphragm, at a mechanical disadvantage and impairs their force-generating capacity
- Consequently, the metabolic work of breathing increases, and dyspnea increases

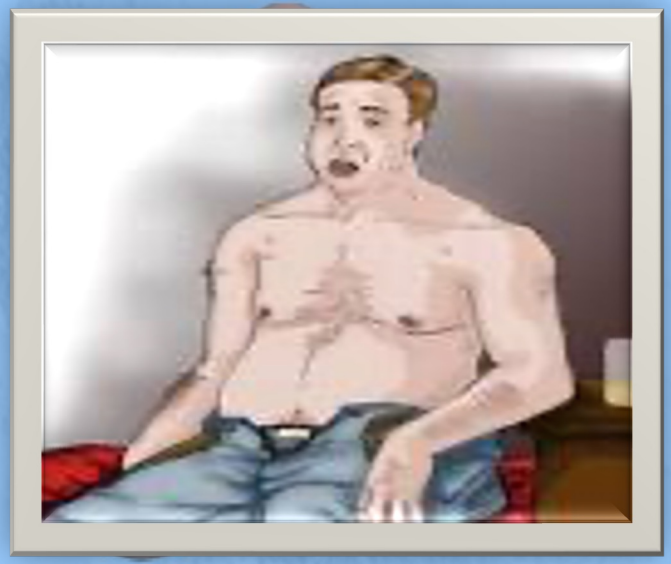


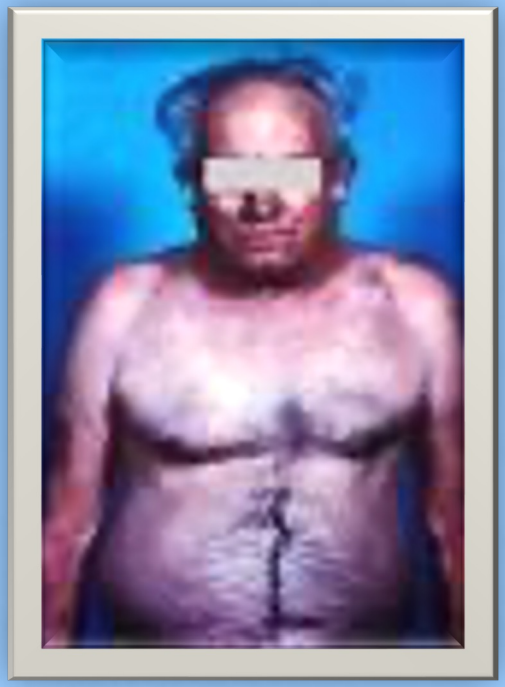

Chronic Bronchitis
Normal
Inflammation and
Structural Changes
Increased
Mucus
Bronchus
Bronchiole
Emphysema
Normal
Bronchiole
Alveoli
Destruction
and Enlargement
of Air Spaces
C. Lynn

- **Two types of COPD**
  - **Type A – Pink Puffers**
    - Have mostly emphysema
    - Need to breathe rapidly to exchange O_2 and CO_2
    - Have prominent dyspnea, the fast puffing keeps them from becoming cyanotic
    - Most of the lung is perfused with blood exchange is not efficient because of fewer alveoli

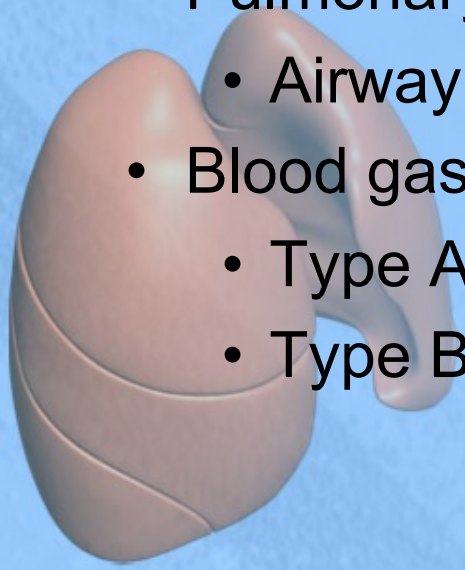


- **Two types of COPD**
  - **Type B – Blue Bloaters**
    - Have mostly chronic bronchitis with bronchiolar obstruction and non-ventilated alveoli
    - Results in shunting of cyanotic blood away from the area where there is no air in the lungs
    - Results in pulmonary hypertension which leads to heart failure with peripheral swelling
    - Severe dyspnea with any exertion

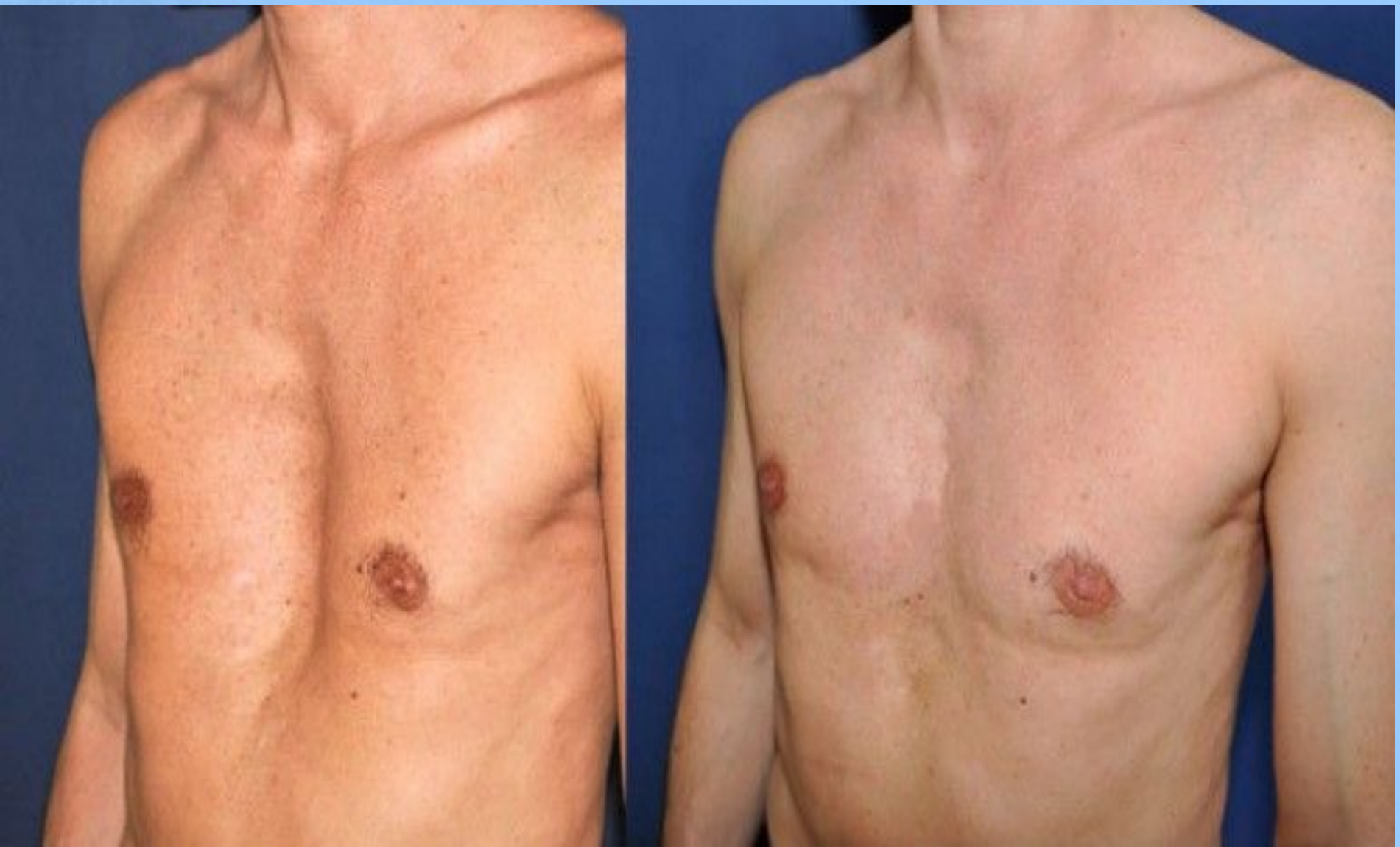




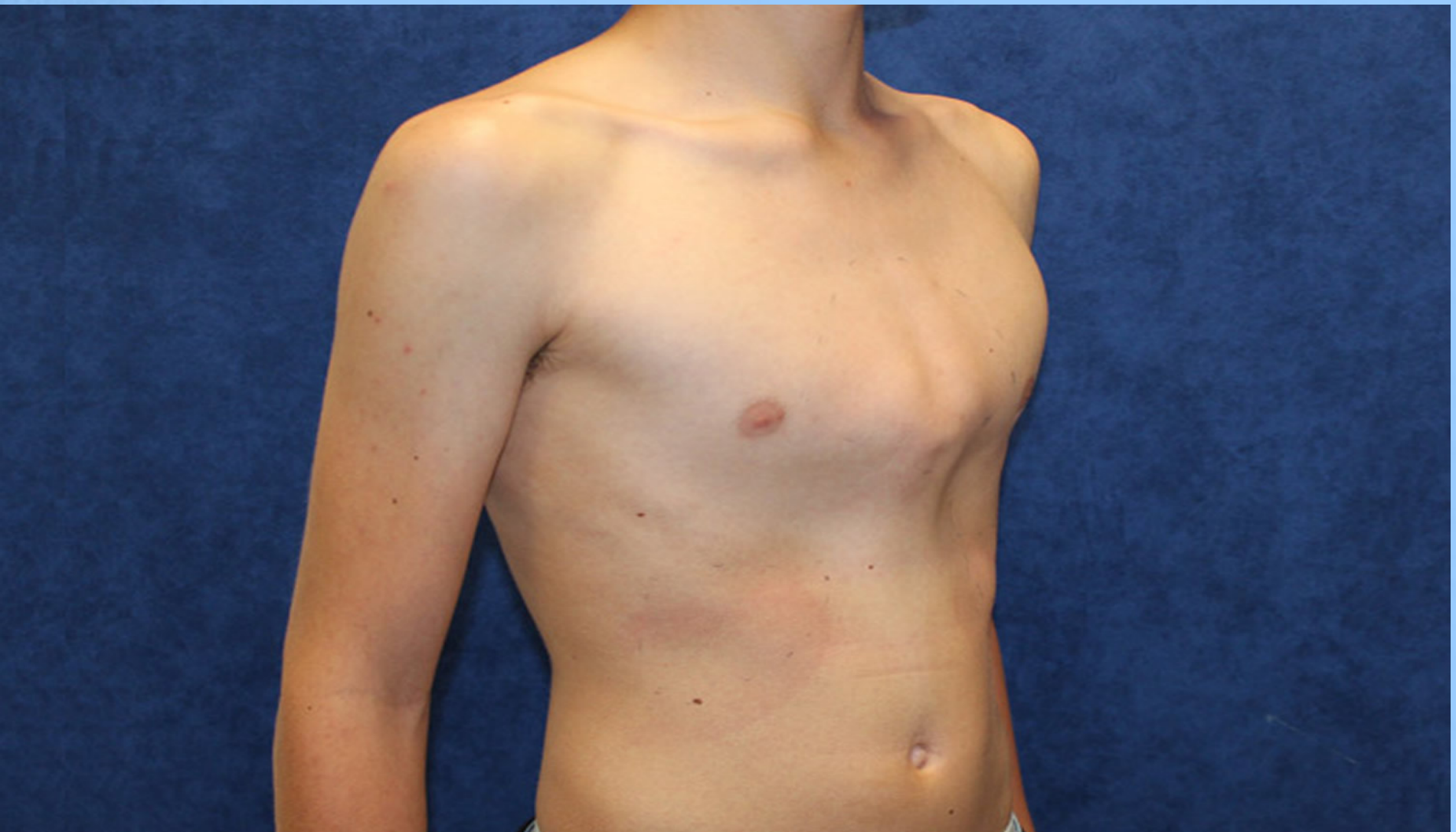
- **Diagnosis**
  - Smoker with hacking cough, sputum and dyspnea
  - Type A – thin, dorsal kyphosis, clubbing, pigeon breast (pectus carinatum) or funnel chest (pectus excavatum)
  - Type B – obese, swollen appearance, cyanotic
  - X-ray findings
    - Large lung volumes hyperlucent, flat diaphragm, increased AP diameter
  - Pulmonary function tests
    - Airway obstruction and decrease, air trapping
  - Blood gases
    - Type A – normal blood gases
    - Type B – marked hypoxemia and CO_2 retention

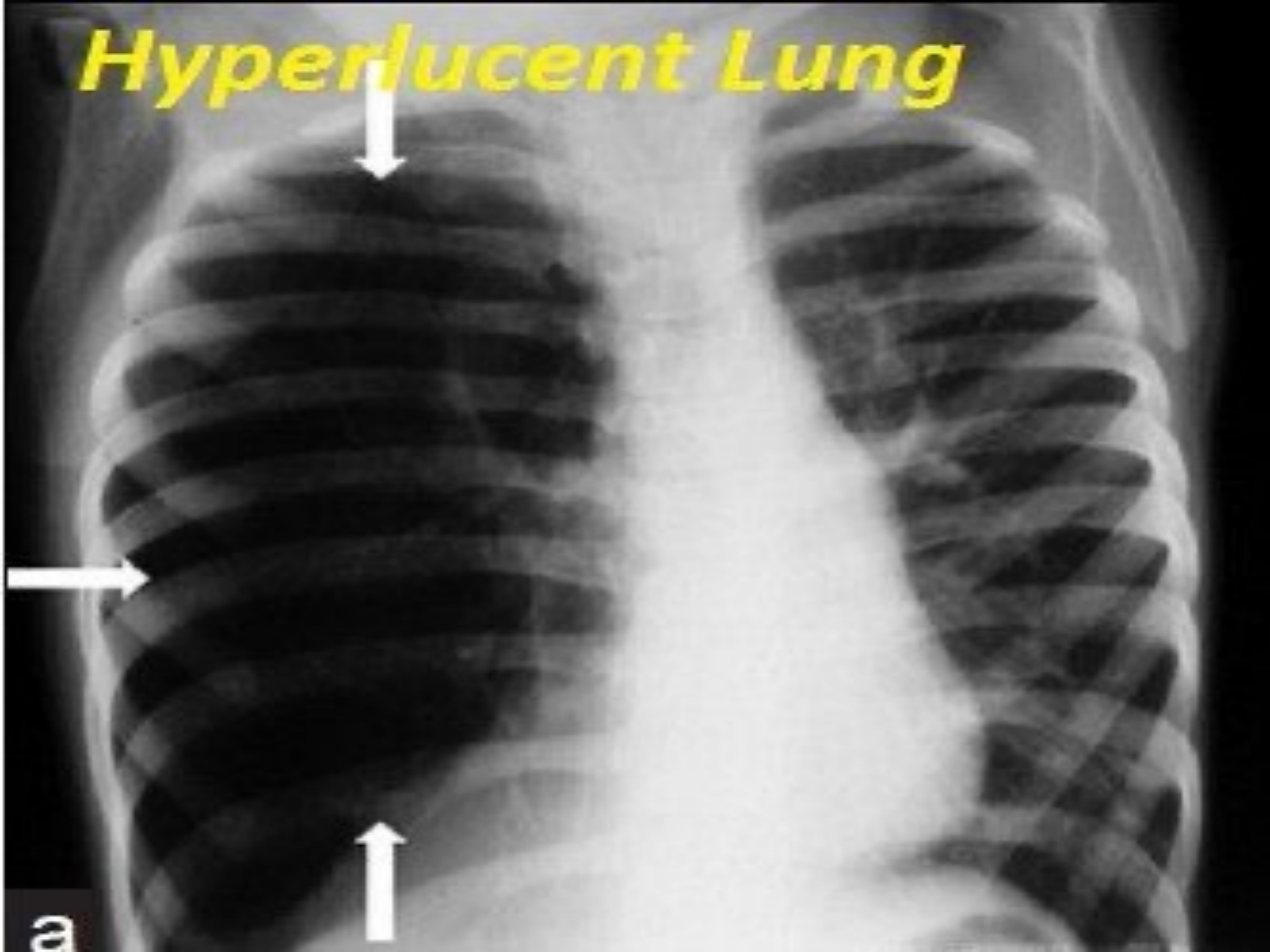


Pectus Excavatum (Funnel Chest)

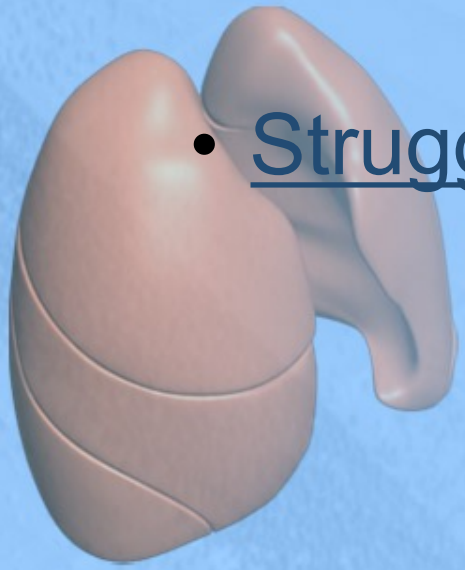


Pectus Carnivatum - Pigeon Chest

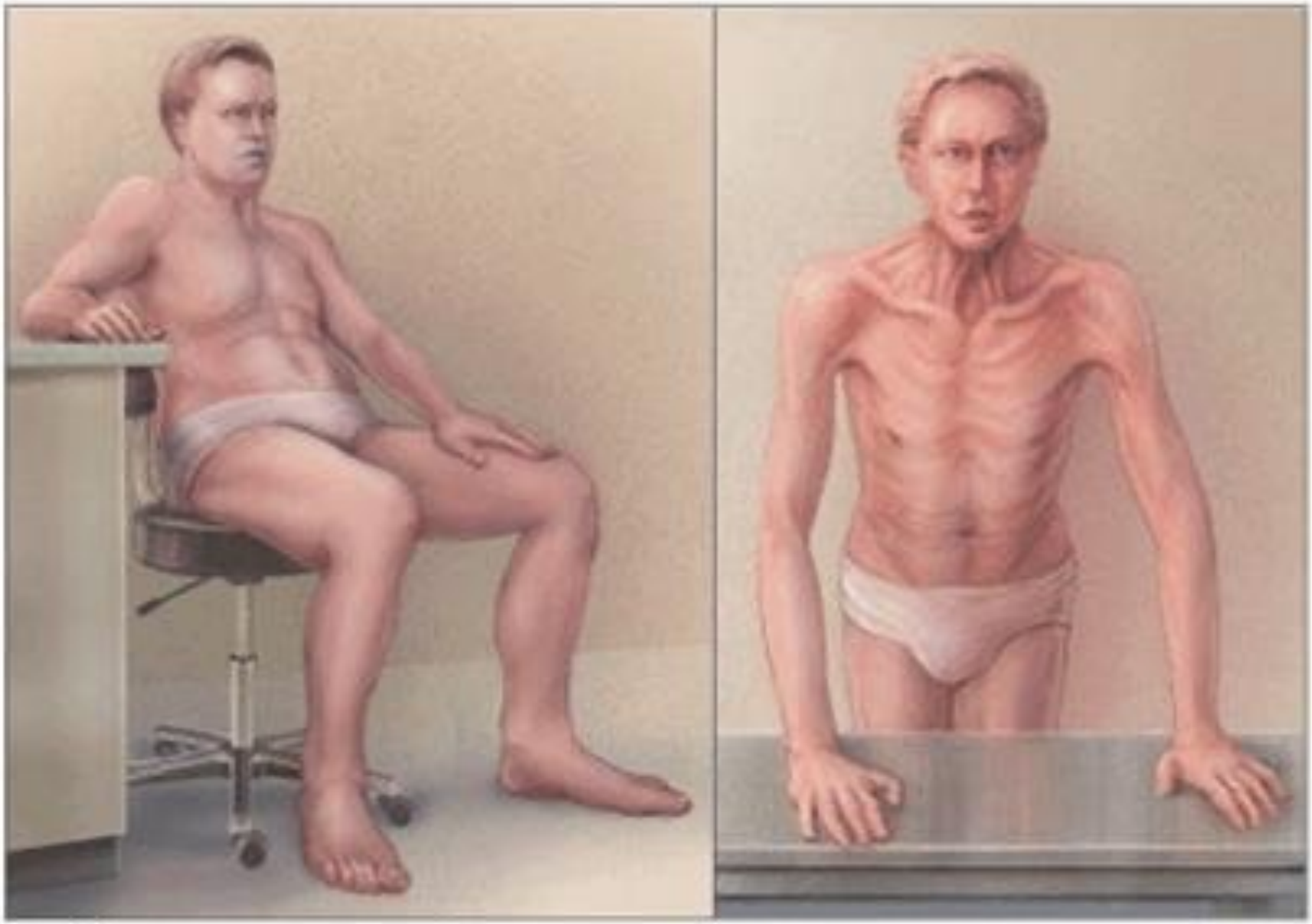


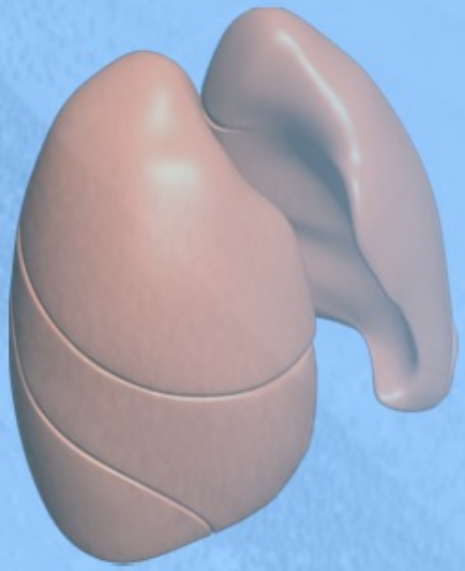

Hyperlucent Lung
a

- Treatment of COPD
  - Bronchodilators
  - Antibiotics
  - Corticosteroids
  - Supplemental oxygen therapy
  - Chest physiotherapy to lose secretions
  - Surgery to remove diseased lung tissue
  - Lung transplantation



- Struggle to Breath

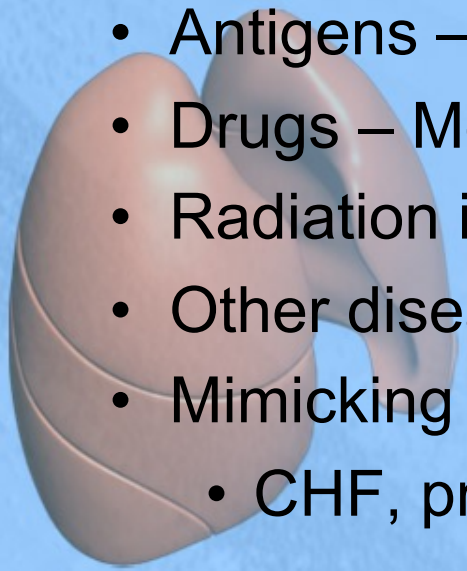




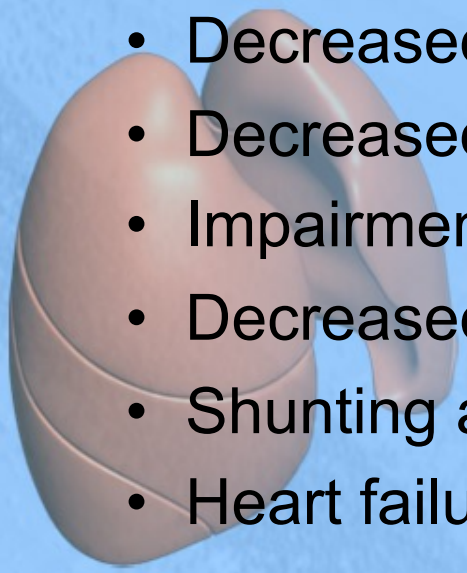
Emphysema puffs pink, chronic bronchitis makes you blue, but no COPD makes me as breathless as you.

Pulmonary fibrosis

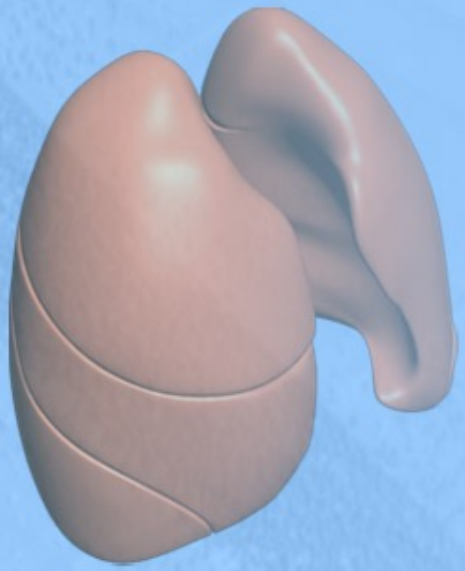
- Referred to as interstitial lung diseases
- Causes inflammation and fibrosis of the connective tissue between the alveoli
- Most common causes
  - Environmental causes – inhaled dusts, asbestosis, silicosis, glass makers, construction workers
  - Antigens – hypersensitivity pneumonitis
  - Drugs – Methotrexate
  - Radiation injury
  - Other diseases – sarcoidosis, RA
  - Mimicking disorders similar presentation but vastly different
    - CHF, pneumocystis or viral pneumonia, carcinomatosis

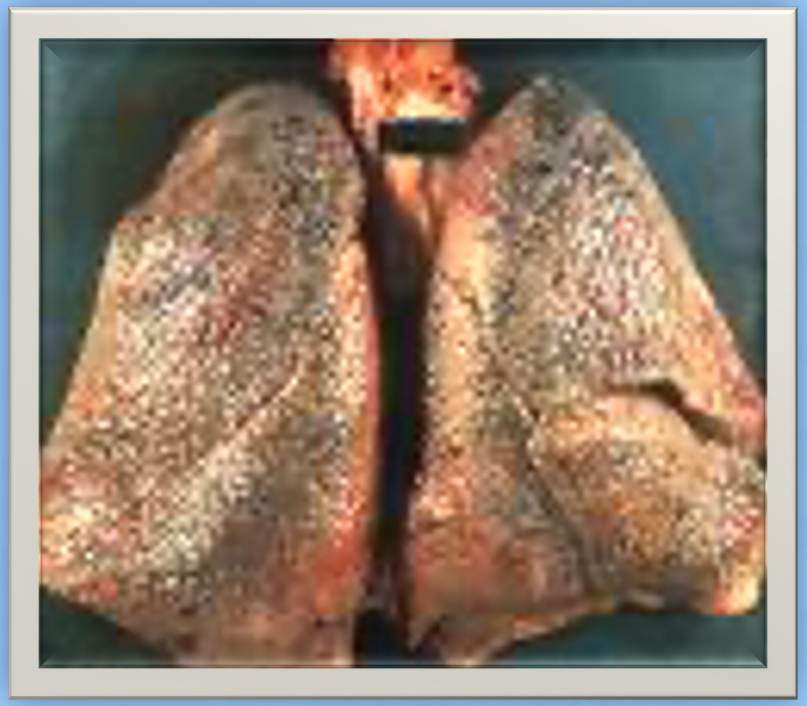


- Pathology of interstitial lung disease
  - Inflammation of the alveolar wall and inter-alveolar spaces
  - Fibrous scarring
  - Granuloma formation
  - End stage leads to a mass of scar tissue with contraction and the formation of cystic areas
- Impairment of pulmonary function
  - Decreased lung volume
  - Decreased compliance (stiff lungs)
  - Impairment of diffusion
  - Decreased gas exchange
  - Shunting and spasm of pulmonary arteries
  - Heart failure resulting from pulmonary hypertension

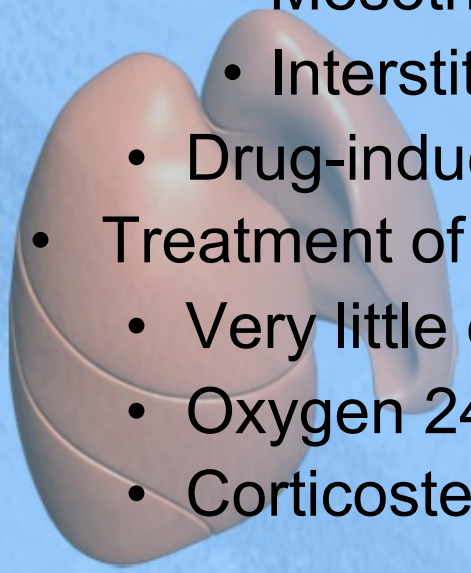


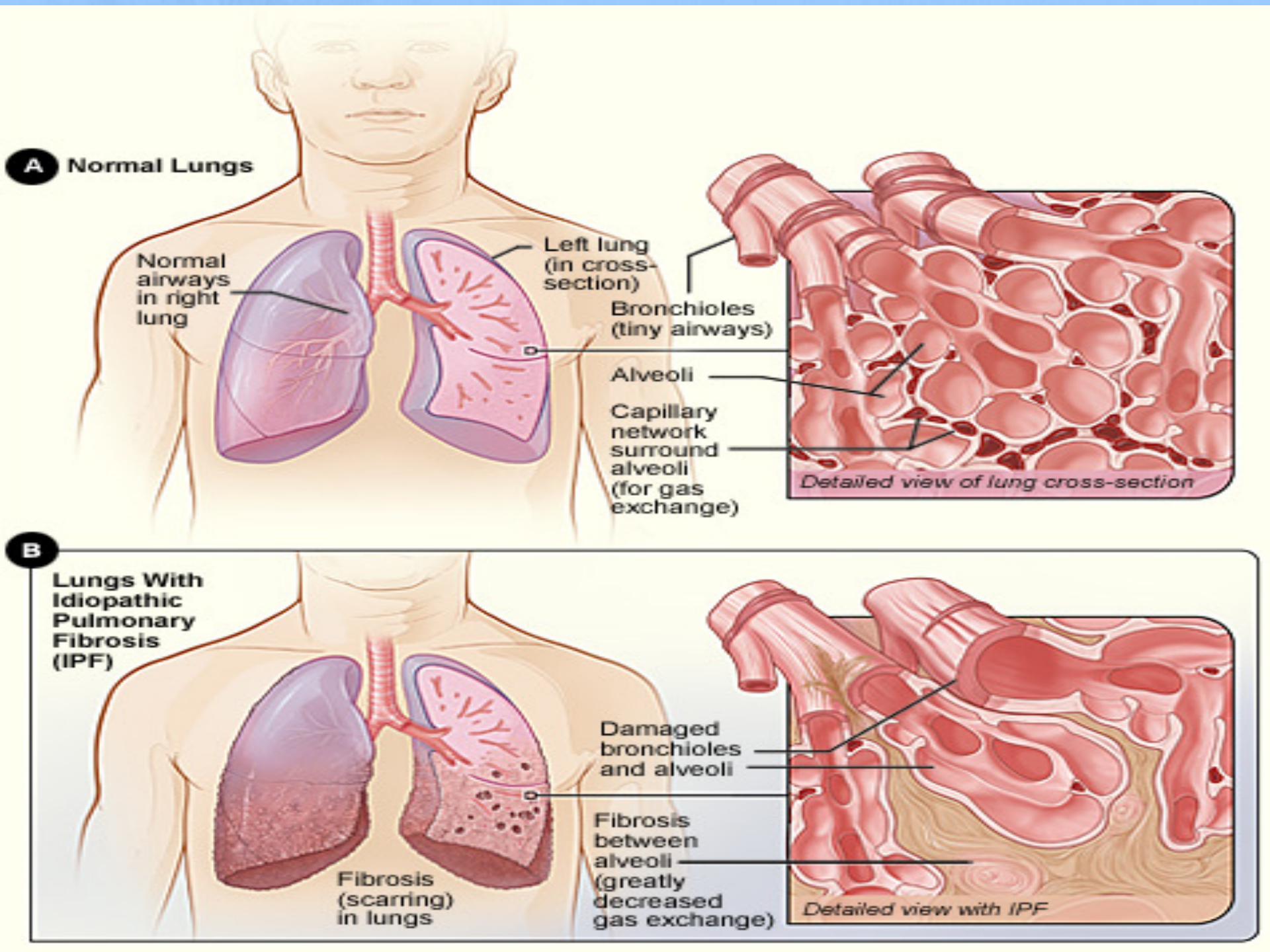
- S & S of pulmonary fibrosis
  - Obvious dyspnea
  - Chronic nonproductive cough
  - Clubbing
  - Mild cyanosis
- Diagnosis of pulmonary fibrosis
  - CT scan is confirmatory





- Specific diseases that can cause pulmonary fibrosis
  - Silicosis – disease of glass makers, sand blasters, rock miners and stone cutters
    - Takes 20 years to develop
  - Pneumoconiosis – coal miner's disease
    - Severe lung fibrosis with hypoxia
  - Asbestosis – leads to 3 distinct diseases
    - Bronchiogenic carcinoma
    - Mesothelioma of lung (cancer of lung pleura)
    - Interstitial fibrosis – takes 20 years to develop
  - Drug-induced pulmonary fibrosis – chemotherapy
- Treatment of pulmonary fibrosis
  - Very little effective care
  - Oxygen 24 / 7
  - Corticosteroids




A Normal Lungs
Normal airways in right lung
Left lung (in cross-section)
Bronchioles (tiny airways)
Alveoli
Capillary network surround alveoli (for gas exchange)
Detailed view of lung cross-section
B
Lungs With Idiopathic Pulmonary Fibrosis (IPF)
Fibrosis (scarring) in lungs
Damaged bronchioles and alveoli
Fibrosis between alveoli (greatly decreased gas exchange)
Detailed view with IPF