DISEASE OF THE RESPIRATORY SYSTEM
Emphysema

- Is characterized by permanent enlargement of the airspaces distal to the terminal bronchioles accompanied by destruction of their walls, without obvious fibrosis.

- Over inflation.

- Types of emphysema:
  1. Centriacinar -> 95%.
  2. Panacinar.
  3. Paraseptal.
  4. Irregular.
Centriacinar (centrilobular) emphysema

• Occur in heavy smoker in association with chronic bronchitis.
• The central or proximal parts of the acini are affected, while distal alveoli are spared.
• More common and severe in the upper lobes, particularly in the apical segments.
• The walls of the emphysematous space contain black pigment.
• Inflammation around bronchi and bronchioles.
Panacinar (panlobular) emphysema

- Acini are uniformly enlarged from the level of the respiratory bronchiole to the terminal blind alveoli.
- More commonly in the lower lung zones.
- Occurs in $\alpha_1$-anti-trypsin deficiency.
Distal acinar (paraseptal) emphysema

- The proximal portion of the acinus is normal but the distal part is dominantly involved.
- Occurs adjacent to areas of fibrosis, scarring or atelectasis.
- More severe in the upper half of the lungs.
- Sometimes forming cyst-like structures with spontaneous pneumothorax.
Emphysema

Incidence

- Emphysema is present in approximately 50% of adults who come to autopsy.
- Pulmonary disease was considered to be responsible for death in 6.5% of these patients.
Irregular Emphysema

- The acinus is irregularly involved, associated with scarring.
- Most common form found in autopsy.
- Asymptomatic.
Pathogenesis

- Is not completely understood.
- Alveolar wall destruction and airspace enlargement invokes excess protease or elastase activity unopposed by appropriate antiprotease regulation (protease-antiprotease hypothesis).
- $\alpha_1$-antitrypsin, normally present in serum, tissue fluids and macrophages, is a major inhibitor of proteases secreted by neutrophils during inflammation.
- Encoded by codominantly expressed genes on the proteinase inhibitor (Pi) locus on chromosome 14.
- The protease-antiprotease hypothesis explain the effect of cigarette smoking in the production of centriacinar emphysema.
  * Smokers have accumulation of neutrophils in their alveoli.
  * Smoking stimulates release of elastase.
  * Smoking enhances elastase activity in macrophages, macrophage elastase is not inhibited by $\alpha_1$-antitrypsin.
**Emphysema**

**Morphology**

- The diagnosis depend largely on the macroscopic appearance of the lung.
- The lungs are pale, voluminous.
- Histologically, thinning and destruction of alveolar walls creating large airspaces.
  - Loss of elastic tissue.
  - Reduced radial traction on the small airways.
  - Alveolar capillaries is diminished.
  - Fibrosis of respiratory bronchioles.
  - Accompanying bronchitis and bronchiolitis.
Emphysema

Clinical course

- Cough and wheezing.
- Weight loss.
- Pulmonary function tests reveal reduced FEV1.

Death from emphysema is related to:
1. Pulmonary failure with respiratory acidosis, hypoxia and coma.
2. Right-sided heart failure.
Other types of emphysema

- Compensatory emphysema.
- Senile emphysema.
- Obstructive overinflation.
- Bullous emphysema.
- Mediastinal (interstitial) emphysema.
## Emphysema and Chronic Bronchitis

<table>
<thead>
<tr>
<th>Appearance</th>
<th>Predominant Bronchitis</th>
<th>Predominant Emphysema</th>
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<tbody>
<tr>
<td>Age</td>
<td>“Blue bloaters”</td>
<td>“Pink Puffers”</td>
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<tr>
<td>Dyspnea</td>
<td>40-45</td>
<td>50-75</td>
</tr>
<tr>
<td>Cough</td>
<td>Mild, late</td>
<td>Severe, early</td>
</tr>
<tr>
<td>Infection</td>
<td>Early, copious sputum</td>
<td>Late, scanty sputum</td>
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<tr>
<td>Respiratory Insufficiency</td>
<td>Common</td>
<td>Occasional</td>
</tr>
<tr>
<td>Cor pulmonale</td>
<td>Common</td>
<td>Terminal</td>
</tr>
<tr>
<td>Airway resistance</td>
<td>Increased</td>
<td>Rare, terminal</td>
</tr>
<tr>
<td>Elastic recoil</td>
<td>Normal</td>
<td>Normal or slightly increased</td>
</tr>
<tr>
<td>Chest radiography</td>
<td>Prominent vessels, large heart</td>
<td>Low</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hyperinflation, small heart</td>
</tr>
</tbody>
</table>
Restrictive Lung Diseases

- Reduced total lung capacity, while the expiratory flow rate is normal or reduced.
- Occur in two general conditions:
  1. Chest wall disorder.
  2. Acute on chronic, interstitial and infiltrative diseases, e.g. ARDS, pneumoconiosis.
Atelectasis

• Incomplete expansion of the lungs or collapse of previously inflated lung substance.
• Significant atelectasis reduce oxygenation and predispose to infection.
Types of Atelectasis

1. Resorption atelectasis.
2. Compression atelectasis.
3. Microatelectasis (patchy atelectasis).
4. Contraction atelectasis.
Types of Atelectasis

1. **Resorption atelectasis.**

   - Result from complete obstruction of an airway and absorption of entrapped air. Obstruction can be caused by:
     
     a. Mucous plug (postoperatively or exudates within small bronchi seen in bronchial asthma and chronic bronchitis).
     b. Aspiration of foreign body.
     c. Neoplasm.
     d. enlarged lymph node

   - The involvement of lung depend on the level of airway obstruction.
   - Lung volume is diminished and the mediastinum may shift toward the atelectatic lung.
Types of atelectasis

2. **Compression atelectasis**

   Results whenever the pleural cavity is partially or completely filled by fluid, blood, tumor or air, e.g.
   - patient with cardiac failure
   - patient with neoplastic effusion
   - patient with abnormal elevation of diaphragm in peritonitis or subdiaphragmatic abscess.
Types of atelectasis

3. Microatelectasis (patchy atelectasis).

• Develops when there is loss of pulmonary surfactant.
• Occur in neonatal or adult respiratory distress syndrome, interstitial inflammation and after surgery.
Types of atelectasis

4. **Contraction atelectasis.**

- Local or generalized fibrotic changes in pleura or lung preventing full expansion of the lung.
Atelectasis

• Atelectatic lung is prone to develop superimposed infection.

• It is reversible disorder except for contraction atelectasis.
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