Restrictive lung diseases. Pneumonia, pleural effusion, pneumothorax, interstitial pulmonary fibrosis (pneumoconiosis, granulomatous and collagenic diseases, drug-induced, idiopathic)

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Restrictive Lung Disease vs Obstructive Problem

• When we are talking about the restrictive lung disease, it is always related to the obstructive lung disease because they are related to the breathing system – although they are different.
  • they may share similar symptoms: shortness of breath, exertional dyspnoe.
  • the diagnose and treatments can be different – depending on patient’s condition, really.

• In obstructive lung disease, there is a difficulty to exhale the air from the lungs because of airways narrowing or lungs damage. The exhaled air will come out slower than the usual, leaving quite a high amount of remaining air within the lungs. This issue is caused by cystic fibrosis, bronchiectasis, asthma, and chronic bronchitis and emphysema.

• The restrictive lung disease is the condition where the lungs can’t be fully expanded
Pathophysiological background of restrictive lung diseases

• Damage of the alveolar walls with three main phases of reaction in the lung
  • Hyaline membranes in alveolar sacks
    • Exudation of fibrin-reach edema fluid with the cytoplasmatic and lipid remnants of necrotic epithelial cells
    • Edema and inflammation of the interstitium
    • Fibrosis in the interstitium

• Two main clinical patterns – depends on which phase of diffuse alveolar damage is most evident
  • Acute restrictive lung disease
    • Exudation and edema
  • Chronic restrictive lung disease
    • Inflammation and fibrosis
Signs and symptoms of interstitial lung diseases

• Dyspnoe
• Hypoxia
• Cor pulmonale
• Clubbing nail
• Cyanosis
• Diffuse infiltration of the lung by small nodules, irregular lines, or „ground-glass“ shadows
Pleural effusion syndrome

Normally, the two (parietal and visceral) sheets of the pleura

- are in close contact (virtual space)
- are moving smoothly over each other

If the surface of the pleural sheets become rough because of apposition of

- fibrin (pleurisy)
- cells (tumour)
- blood clot (injury)
- callous tissue (tbc)

then each breath will cause pleural rub
Pleural rub

• Normal pleural surfaces move smoothly and noiselessly against each other during respiration.

• When pleural surfaces become inflamed, they move jerkily as they are repeatedly delayed by increased friction.

• The sounds may be discrete, but sometimes are so numerous that they merge into an apparently continuous sound. It is localized to a relatively small area of the chest wall.
Pleural effusion

• Pleural fluid accumulates when pleural fluid formation exceeds pleural fluid absorption

• Fluid enters the pleural space:
  • from the capillary in the parietal pleura (removed lymphatics of parietal pleura)
  • from the interstitial spaces of the lung via the visceral pleura
  • From the peritoneal cavity via small holes of diaphragm
To determine whether the effusion is transudate or exudate

- Transudative pleural effusion occurs when **systemic factor** that influence the formation and absorption of pleural fluid altered
- Exudative pleural effusion occurs when **local factor** that influence the formation and absorption of pleural fluid altered
- Exudative pleural effusions meet at least one of the following criteria:
  - Pleural fluid/serum protein >0.5
  - Pleural fluid/serum LDH >0.6
  - Pleural fluid LDH more than two-thirds of the normal upper limit for serum
### Causes of pleural effusion

<table>
<thead>
<tr>
<th>Transudate</th>
<th>Exudate</th>
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<tbody>
<tr>
<td><strong>Congestive heart failure</strong></td>
<td>Infections</td>
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<tr>
<td><strong>Liver cirrhosis</strong></td>
<td>Tumours (lung, breast, lymphoma)</td>
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<tr>
<td>Nephrotic syndrome</td>
<td>Pulmonary embolism</td>
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<tr>
<td>Peritoneal dialysis</td>
<td>Vasculitis – autoimmune disorders</td>
</tr>
<tr>
<td>Myxoedema</td>
<td>Empyema thoracis</td>
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<tr>
<td><strong>Meigs’s syndrome (benign ovarian tumours can cause ascites and a pleural effusion)</strong></td>
<td>Tuberculous pleuritis</td>
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<td>After irradiiation</td>
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<td></td>
<td>Gastrointestinal disorders (pancreatitis)</td>
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<tr>
<td></td>
<td>Hemothorax (iatrogenic or traumatic)</td>
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</table>
Pleural effusion syndrome, physical signs

- **Respiratory expansion:**
  - decreased

- **Palpation:**
  - pectoral fremitus is decreased

- **Percussion:**
  - **Dullnes.** Ellis-Damoiseau's line, Korányi-Grocco-Rauchfuss triangle. Borders are different if:
    - air gets over the fluid (pleuro-px)
    - fluid is trapped by callus

- **Auscultation:**
  - breathing sounds are decreased. However: compression of adjacent lung tissue causes atelectasis → fine crackles and bronchial breathing
• **Assessment of respiratory expansion:**
  
  • Place your thumbs about at the level of and parallel to the 10th ribs, your hand grasping the lateral rib cage
  
  • Slide your hands medially a bit in order to raise loose skin folds between thumbs and spine
  
  • Ask the patient to inhale deeply
  
  • During inhalation the chest expands, and the thumbs move away from the spine
  
  • Watch the divergence of your thumbs during inspiration and feel for the range and symmetry of respiratory movement
Tactile fremitus – refers to the palpable vibrations transmitted through the bronchopulmonary system to the chest wall when the patient speaks

• Palpate and compare symmetrical areas of the lungs
• Use the ulnar surface of your hand. Ask the patient to repeat ninety-nine or „harminchárom”.
• If fremitus is faint, ask the patient to speak more loudly or in a deeper voice.
X-ray in pleural effusion syndrome

Small amount of pleural effusion

Large amount of pleural effusion
Pneumothorax syndrome I.

• When air leaks into the pleural space:
  • **Primary spontaneous ptx:** usually due to rupture of apical pleural bulla. Almost exclusively in smokers.
  • **Secondary ptx:** due to chronic obstructive lung disease.
  • **Traumatic ptx:** penetrating or non-penetrating, iatrogenic (insertion of central intravenous catheters)
  • **Tensile ptx:** usually occurs during mechanical ventilation or resuscitative efforts. The positive pleural pressure is life-threatening:
    • Severely compromised ventilation
    • Decreased venous return to the heart and reduced cardiac output
Pneumothorax syndrome II.

1. **Decreased** chest expansion
2. Heart and mediastinum is **dislocated** toward the opposite side
3. Hyperresonant or **tympanitic** percussion sound
4. Decreased or **absent breathing sounds**
5. **Decreased** or absent **tactile fremitus**
6. Sudden pain, dyspnoea, cyanosis
X-ray picture of pneumothorax
Pulmonary infiltration syndrome

The normally air-filled lung contains an area where:

- alveoli are filled by fluid
  - exudate (pneumonia)
  - transudate (congestive heart failure)
  - blood (pulmonary embolism)
- normal tissue is replaced by solid tumour
- alveoli are compressed (around a tumour)
  - air is reabsorbed from the alveoli, thus they collapse (atelectasis syndrome)
Pulmonary infiltration syndrome

The infiltration **may respect** the borders of
- lung lobes (lobar pneumonia)
- lung segments (embolism, atelectasis)

or **may not respect** the borders:
- tumour
- bronchopneumonia
- congestion
Nosocomial pneumonia

- accounts for 15-20% of nosocomial infections
- are caused by aspiration of endogenous or hospital-acquired oropharyngeal (and occasionally gastric) flora
- are associated with more deaths than are infections at any other body site
- Risk factors include those events that increase colonization by potential pathogen: prior antimicrobial therapy, contaminated ventilator circuits, decreased gastric acidity, intubation, nasogastric tube etc.
- The most likely pathogens:
  - Early-onset nosocomial pneumonia (within 4 days after admission): Streptococcus pneumoniae and Haemophilus species
  - Late-onset nosocomial pneumonia: S. aureus, Pseudomonas aeruginosa, Enterobacter spec., Klebsiella pneumoniae, Acinetobacter baumani
Cases of atypical pneumonia do not usually require hospitalization, and a person with it is unlikely to be significantly ill. This is why it is often called „walking pneumonia“.

With atypical pneumonia will also have certain extrapulmonary symptoms that others with typical pneumonia will often not have.

Atypical pneumonia is caused by atypical bacteria that do not stain with Gram stain.

Despite general symptoms and problems there are in general few physical signs. The patient looks better than the symptoms suggest.

Absence of leukocytosis in atypical pneumonia.

Symptoms of atypical pneumonia tend to be milder and more persistent than those of typical pneumonia.

Atypical pneumonia requires different antibiotic than typical pneumonia.
Characteristics of community acquired pneumonia

• **Typical pneumonia**
  - Acute onset
  - Fever, chills
  - Productive cough
  - Pleural pain
  - Physical signs (+)
  - Lobar consolidation

• **Agents**
  - Streptococcus pneumoniae
  - Haemophylus influenzae
  - Gr – bacillus
  - Anaerobes

• **Atypical pneumonia**
  - Insidious onset
  - Protracted clinical course
  - Subfebrility
  - Non-productive cough
  - Nonrespiratory symptoms
  - Physical signs (-)
  - Non-lobar infiltration

• **Agents**
  - Mycoplasma pneumoniae
  - Chlamydia pneumoniae
  - Legionella pneumophila
  - Virus
Streptococcal pneumonia

- Leading cause of community acquired pneumonia, particularly in young and old patients
- Abrupt onset of chills
- Continua fever (without antibiotic treatment)
- Chest pain
- Rust colored sputum
- Treatment: resistant strains have appeared
- Prevention: immunization
Special characteristics of other bacterial pneumonias

• Staphylococcus pneumonia: multifocal, bilateral. It may cause cavitating pneumonia with pleural effusion.


• Moraxella catarrhalis: In older adults with COPD or in immunocompromised patients.
<table>
<thead>
<tr>
<th>Agent</th>
<th>Epidemiology (source, widespread)</th>
<th>Pulmonary signs</th>
<th>Systemic symptoms</th>
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<tbody>
<tr>
<td>Legionella pneumophila</td>
<td>Flagellated, non-spore forming Gram negative</td>
<td>Non-productive cough, high fever, dyspnoe</td>
<td>Confusion, diarrhea, hyponatremia, hepatic dysfunction, bradycardia</td>
</tr>
<tr>
<td>Mycoplasma pneumoniae</td>
<td>Smallest free-living organism that lacks a cell wall and cannot be stained</td>
<td>Sore throat, persistant, slowly worsening dry cough. Multifocal bilateral diffuse infiltrates, pleural effusion is not rare</td>
<td>Splenomegaly, lymphadenopathy, maculopapular skin rash, bullous myringitis</td>
</tr>
<tr>
<td>Chlamydia pneumoniae</td>
<td>Intracellular</td>
<td>Prolonged sore throat and hoarseness. Gradual onset of cough with little or no fever. Subsegmental infiltrate</td>
<td></td>
</tr>
<tr>
<td>Q-fever</td>
<td>Coxiella burnetti Gram negative intracellular bacterium</td>
<td>50%- flu-like symptoms, Pneumonia with mild cough, self-limited disease.</td>
<td>Granulomatous hepatitis, Chronic disease in patients with pre-existing heart condition - endocarditis</td>
</tr>
</tbody>
</table>
Chest X-ray in pulmonary infiltration syndrome

Lobar pneumonia

Bronchopneumonia
Pulmonary infiltration syndrome III.

- **Respiratory expansion:**
  - diminished

- **Palpation:** tactile (pectoral) fremitus
  - increased

- **Trachea:**
  - always in the midline

- **Percussion:**
  - **dullness** over the affected area (may be absent in deep-sited lesions)

- **Auscultation:**
  - **fine crackles** (crepitation) if alveoli are filled with fluid (pneumonia, embolism, congestion) or partly compressed (tumour), but they are absent if the alveoli are airless (atelectasis)
  - **bronchial breathing** sound
• Discontinuous sounds (crackles, rales)
  • short intermittent
  • non-musical
  • typically inspiratory sound
  • they result from a series of tiny explosions when small airways deflated during expiration, pop open during inspiration.
  • it can be simulated by rolling a lock of hair between your fingers close to the ear
  • Fine crackles: produced in the alveoli (late inspiratory, repeat themselves from breath to breath) Coarse crackles (early inspiratory) : arise in the bronchioli
Extrinsic allergic alveolitis
(hypersensitivity pneumonitis)

• The hypersensitivity reaction in the lung occurs in response to inhaled organic dust
• The exposure may be occupational or environmental
• The disease occurs from type III and IV hypersensitivity reactions
• The alveolar walls are infiltrated by lymphocytes, histiocytes and plasma cells.
• There are loosely formed granulomas
• Fibrotic changes occur in advanced disease
Etiologic agents in hypersensitivity pneumonitis

- Farmer’s lung – moldy hay - thermoactinomyces vulgaris
- Malt worker’s lung- moldy malt- aspergillus clavatus
- Cheese worker’s lung- cheese mold – penicillium caseii
- Bird breeder’s lung- pigeon, parakeets, fowl – avian or animal proteins (in secrete)
• Acute form
  • Dyspnea, fever, malaise and cough appear 4-6 hours after exposure and continue for 24-48 hours.
    • Fine crackles throughout the lungs.
    • Chest radiograph: may be normal, but may show reticular nodular infiltration

• Chronic form
  • Progressive dyspnea
  • Hypoxemia which worsens on exercise
  • Bilateral inspiratory crackles
  • Chest radiograph: reticular nodular infiltration, fibrosis predominantly in upper lobes
  • Pulmonary function tests: restrictive pattern
Asbestosis

• Asbestosis is a chronic lung disease. This disease is caused when the asbestos fibers are inhaled.
• Asbestos is a natural mineral product. This natural mineral product is resistant to corrosion and heat. In the past this product has been used in products such as cement, floor tiles and insulation.
• Many of the people who are suffering from asbestosis have got this disease from their jobs.
• These types of jobs will make increased chances to get this disease:
  • Boiler operators
  • Electricians
  • Auto mechanics
  • Aircraft mechanics
  • Railroad workers
  • Asbestos miners
  • Building construction workers
  • Workers which are removing the asbestos insulation around steam pipes in the older buildings
Clinical features of asbestosis

• After inhalation of this dust, microscopic asbestos fibers are deposited in the lungs where they can cause permanent damage, in addition to chronic respiratory symptoms.

• Long latency period between exposure to asbestos and the onset of the resulting disease. For example, 1 year of exposure during youth can cause symptoms that will only appear 30 years later.

• Respiratory problems worsen progressively and in about 15% of cases, severe breathlessness and respiratory failure are manifested.

• The risk of lung cancer is considerably higher for a person who smokes and has been exposed to asbestos.

• Pulmonary transplantation remains the only way to treat end-stage asbestosis.
Sarcoidosis

• Characterized by the presence of granulomatous tissue (non-caseating granuloma composed by histiocytes, giant cells, and lymphocytes)

• Systemic disease which involves the eyes, brain, heart, lungs, bones, kidneys, skin, liver and spleen.

• Etiology: Unknown, likely immunological basis

• Incidence: 50/100 000, mostly in young females

• 85% of these patients improve spontaneously, but 15% may develop progressive lung fibrosis and respiratory failure.
Clinical features in sarcoidosis

- **Stage 0**: no obvious intrathoracic involvement. Normal pulmonary function.
- **Stage I**: Bilateral hilar lymphadenopathy, often accompanied by arthritis, uveitis, erythema nodosum. Normal pulmonary function.
  - Associated signs and syndromes: cough, dyspnoe, fever, weight loss, fatigue, hypercalcemia, increased red blood cell sedimentation rate.
- **Stage II**: Pulmonary parenchyma is also involved, changes in mid and upper zones. Restrictive changes in pulmonary function.
- **Stage III**: Pulmonary infiltrates and fibrosis without adenopathy. Restrictive changes in pulmonary function.