Restrictive and obstructive lung disorders

By Maria W. Lied

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   - → Lung volumes
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   - → COPD
3. Restrictive lung disorders
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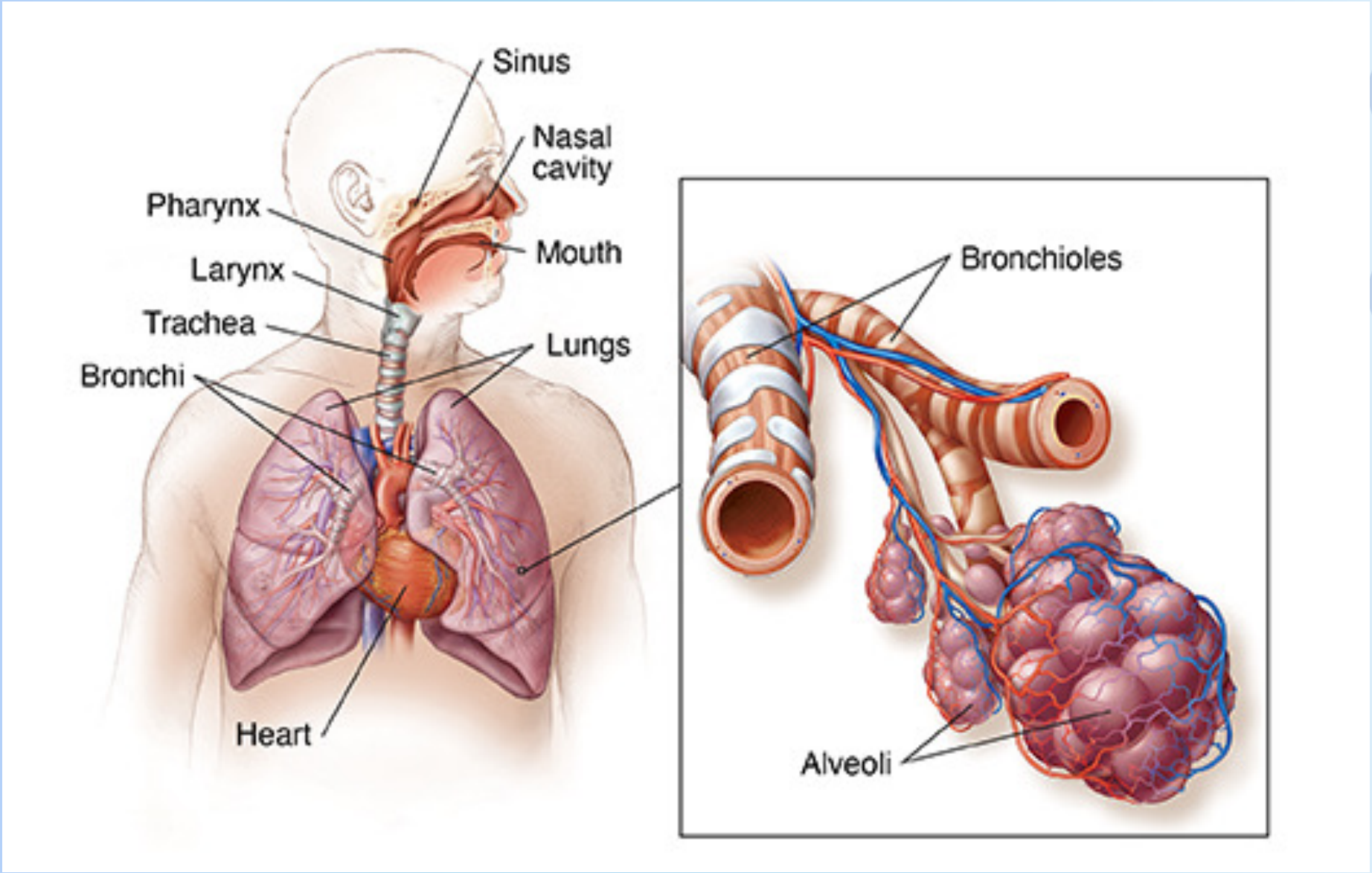
   - → Sarcoidosis


Sinus
Nasal cavity
Pharynx
Mouth
Larynx
Trachea
Lungs
Bronchi
Heart
Bronchioles
Alveoli

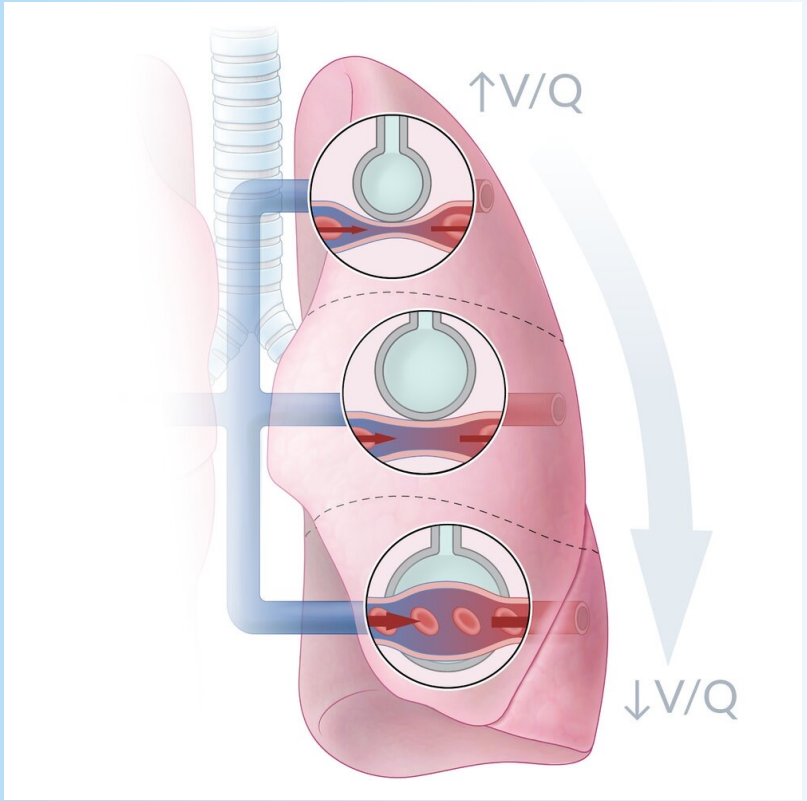
Ventilation and perfusion

Definition

Ventilation (V)
The movement of air between the atmosphere and the lungs

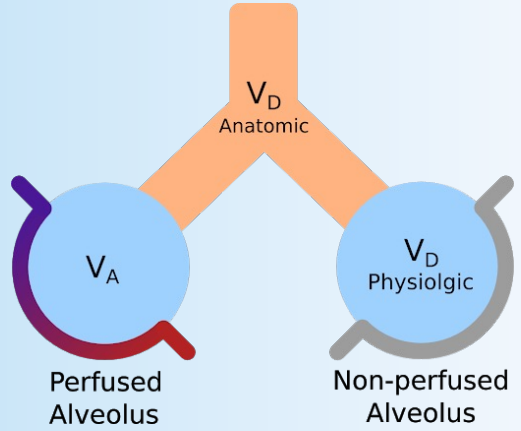
Perfusion (Q)
The delivery of blood to the alveoli

$\frac{V}{Q}$ ratio



Diffusion
Gas exchange

Dead space



Anatomic dead space

The parts of the respiratory system that conduct air but do not participate in gas exchange

Physiologic dead space

The volume of inspired air that does not participate in gas exchange

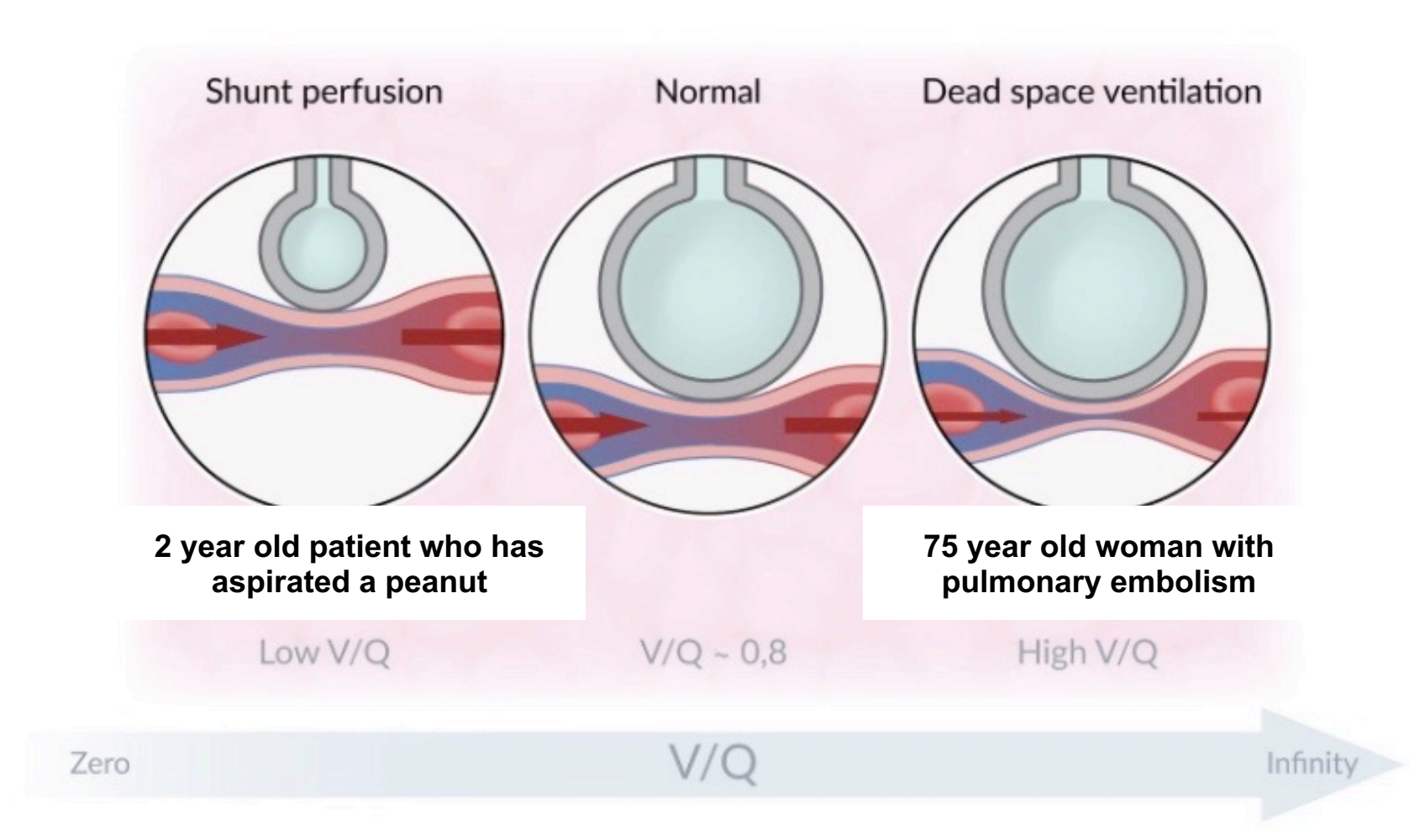
Physiologic conditions

Physiologic dead space = anatomic dead space

Pathologic conditions

Physiologic dead space > anatomic dead space ➔ V/Q mismatch

Ventilation/perfusion mismatch



Spirometry

Forced vital capacity (FVC)

→ Maximum amount of air a person can inhale after a forceful exhalation.

Functional residual capacity (FRC)

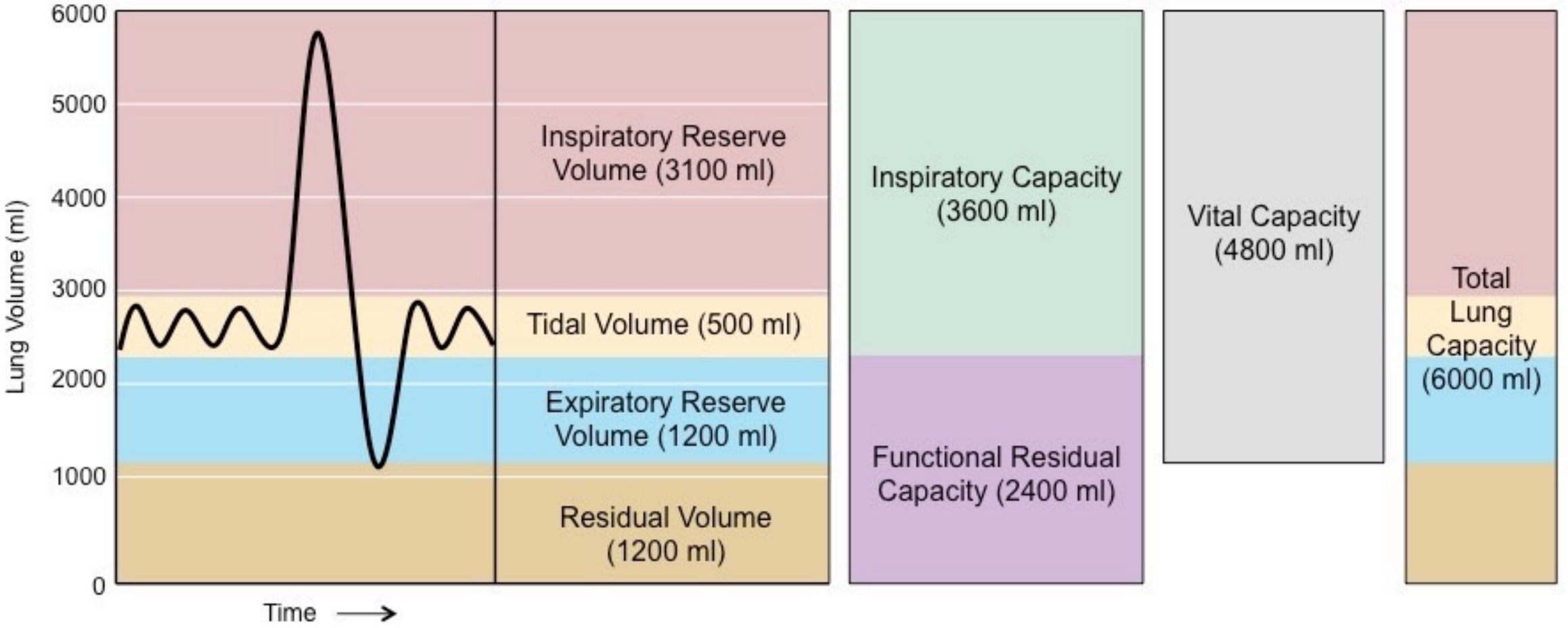
→ Volume remaining in the lungs after a normal, passive exhalation

Total lung capacity (TLC)

→ Volume of air of inhalation and exhalation under volunteer and unvolunteered activity

FEV_1

→ Proportion of a person`s vital capacity that they can expire within the first second of forced expiration.



Obstructive lung disorder

Increased resistance to airflow caused by narrowing/obstruction of airways.

= makes it harder to breathe out

Restrictive lung disorder

Imparied ability for the lungs to expand.

= makes it harder to breathe in

Content

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~~→ Principe of V/Q~~

~~→ Lung volumes~~

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   - → Idiopathic pulmonary fibrosis
   - → Exposure-related restrictive lung diseases
   - → Sarcoidosis

Obstructive lung disorders

→ ASTHMA
→ COPD

Asthma

The most common chronic disorder of childhood

Pathomorphology note:
The mucus of an asthmatic patient contain:
- Curschmann spirals
- Charcot-Leyden crystals

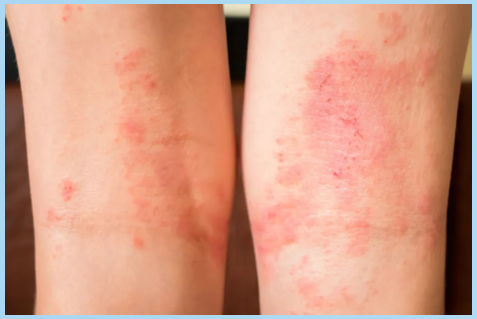
Difficulty to breathe
→ Upper respiratory tract infections
→ In relation to physical activity

Persistent, dry cough

Weezing on expiration

Allergic asthma

- Child
- Eczema
- Family history
- Responsive to ICS



Non-allergic asthma

- Adult
- Smoker
- Construction worker - living in cities with bad air quality
- Obese
- Stress
- Not responsive to ICS

*ICS = inhaled corticosteroids

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Pathogenesis of allergic asthma

A type 1 hypersensitivity reaction (IgE)

1. Bronchial hyperresponsiveness

Triggered by inhalation of an antigen: dust, pollen, pet hair, mold spores
Lead to activation of Th_2-cells which cause release of cytokines

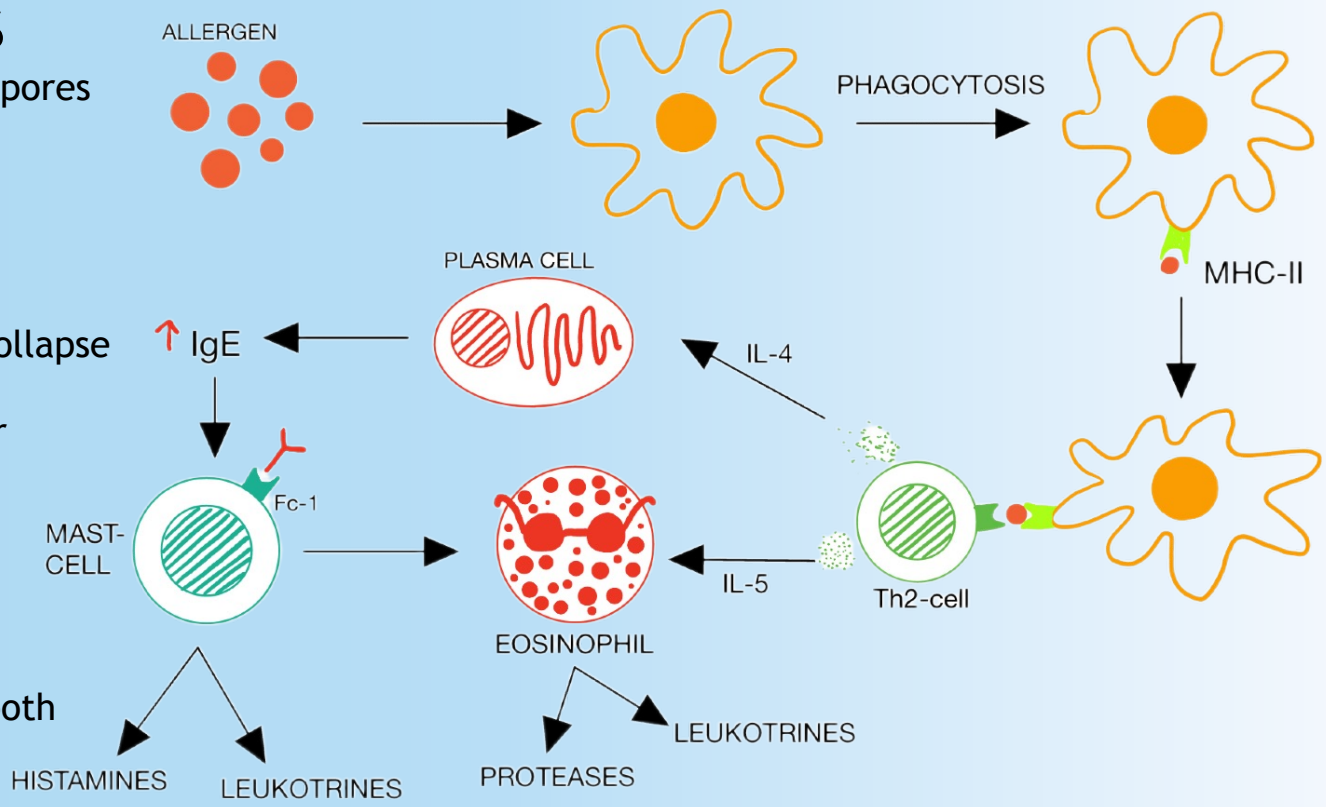
2. Bronchial inflammation

- Edema of bronchioles & smooth muscle contraction → bronchial collapse
- Increased mucus production
- Activation of eosinophils and plasma cells and induction of cellular response: ↑ vascular permeability

3. Endobronchial obstruction

Trouble breathing Out = obstructive lung disorder
Bronchospasm, increased mucus production and hypertrophy of smooth muscle cells
→ REVERSIBLE

Chronic hypertrophy of smooth muscle cells can lead to fibrosis of lung tissue
→ IRREVERSIBLE



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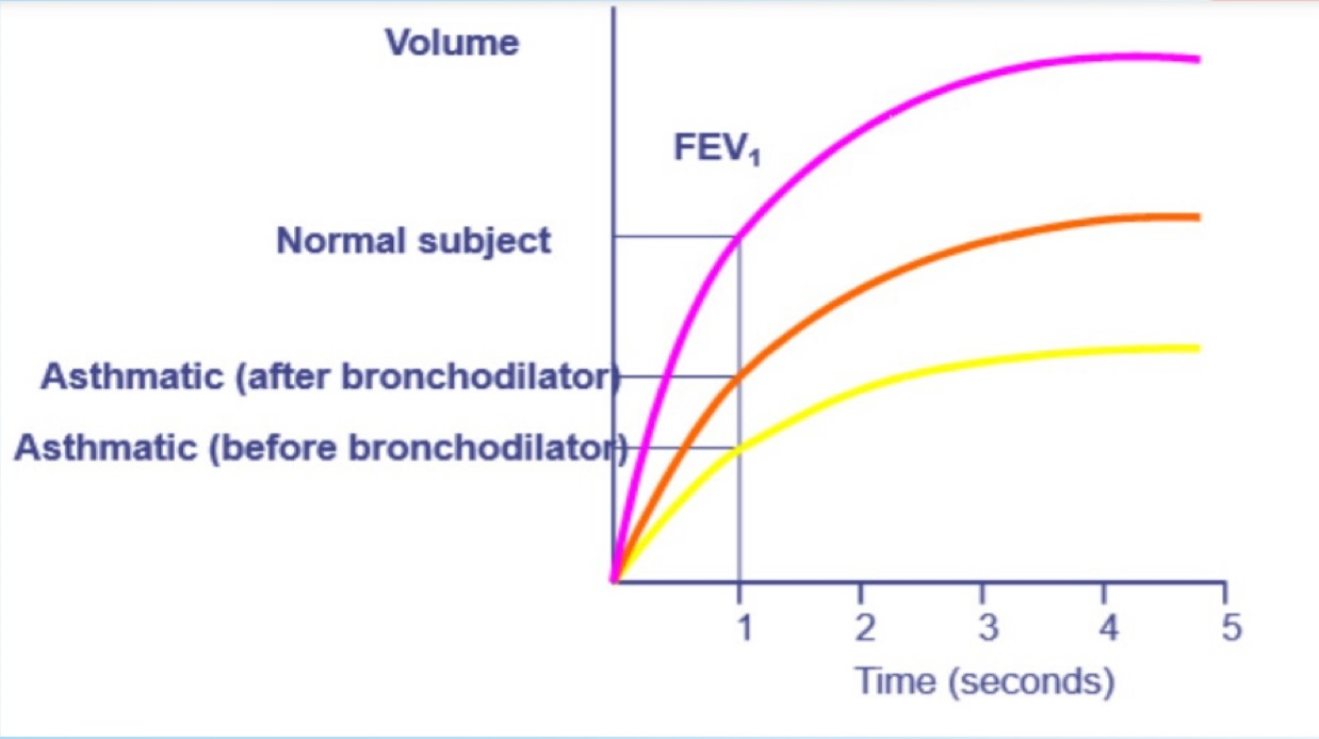
Diagnosis

- Typical clinical features of asthma
- Demonstration of reversible bronchial obstruction

  With the use of bronchodilators.
  Significant improvement: >12% improvement of FEV_1



Asthma can also be partly diagnosed based on measurements of nitric oxide exhaled. Epithelial cells in inflammation of airways produce NO ;))

Chronic obstructive pulmonary disorder

COPD

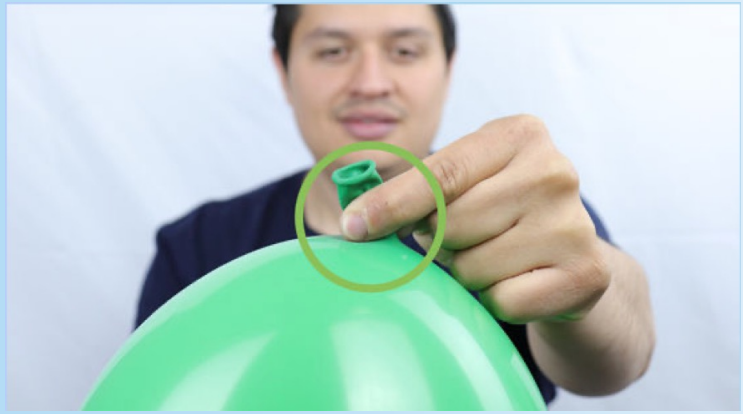
→ Bronchitis
→ Emphysema

Key facts about COPD

- Divided into chronic bronchitis and emphysema.
- Third leading cause of death.
- Characterized by irreversible airflow obstruction due to chronic inflammation of the small airways and parenchymal destruction.

Risk factors:

- Smoking (90%)
- Air pollution
- Upper respiratory tract infections
- α-1 antitrypsin deficiency (emphysema)



UWAGA:
Patients usually present with both chronic bronchitis AND emphysema, the distinction of the 2 diseases is mainly for exams 😉

Pathogenesis - chronic bronchitis

1. Long-time exposure of irritant

2. Hyperplasia and hypertrophy of mucus glands

Bronchial mucus cells in bronchi

Goblet cells in bronchioles

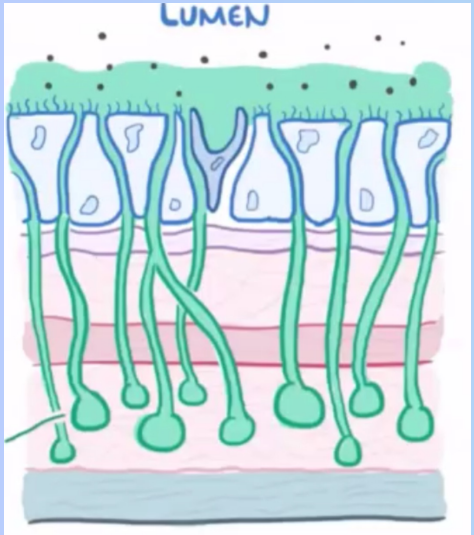
3. CO_2 build up

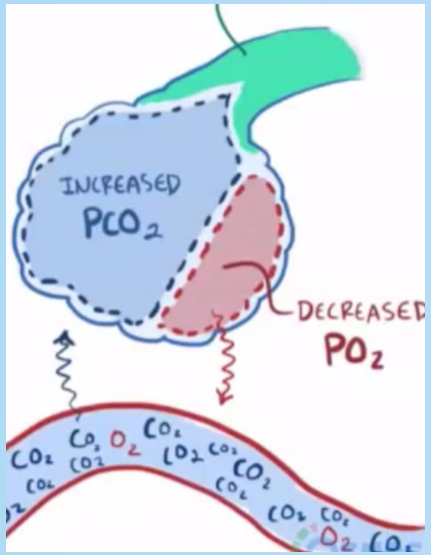
4. Hypercapnia and hypoxemia

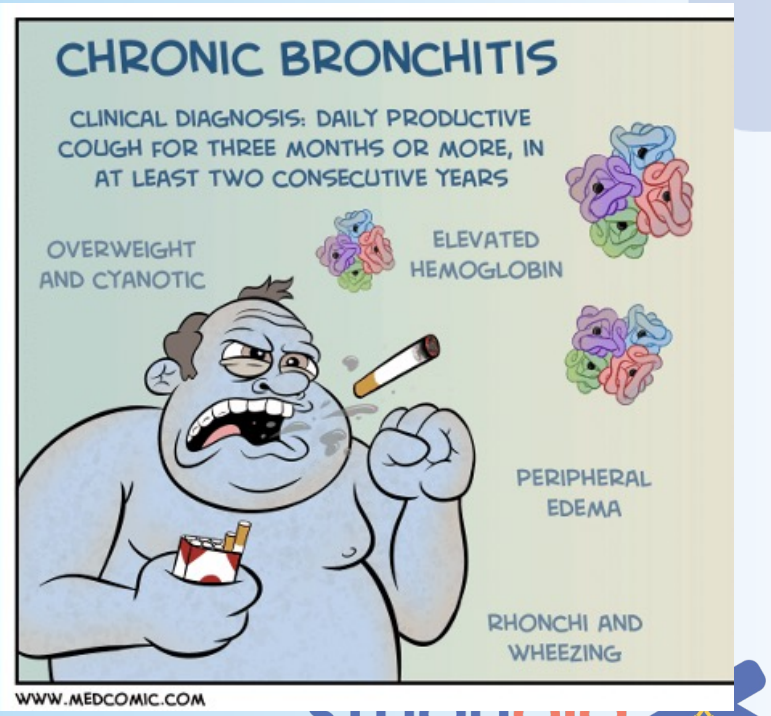
Hypercapnia: An elevation in arterial CO_2-levels

Hypoxemia: The below-normal level of oxygen in your blood, specially in arteries.

"The blue bloater"







Pathogenesis - Emphysema

α-1 antitrypsin deficiency

1. Long-time exposure to irritants

2. Hypersensitivity reaction

Activation of neutrophils

Release of proteases (elastase)

3. Elastase break down elastin

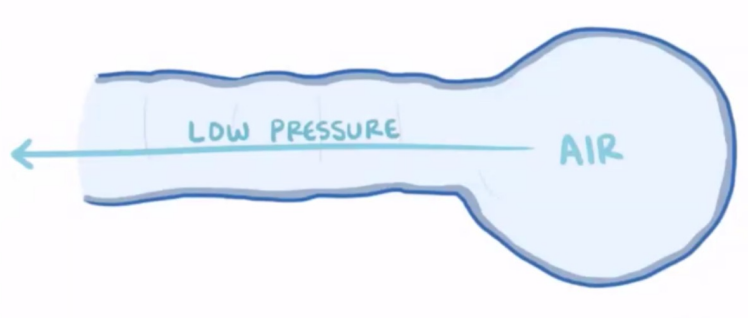
Elastin is crutial for elastic recoil

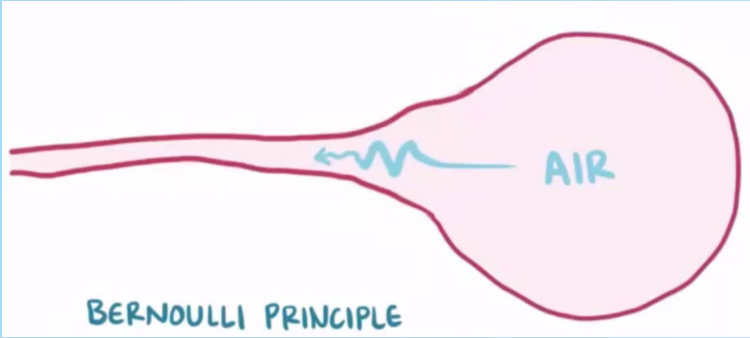
No elastin = collapse of bronchiole

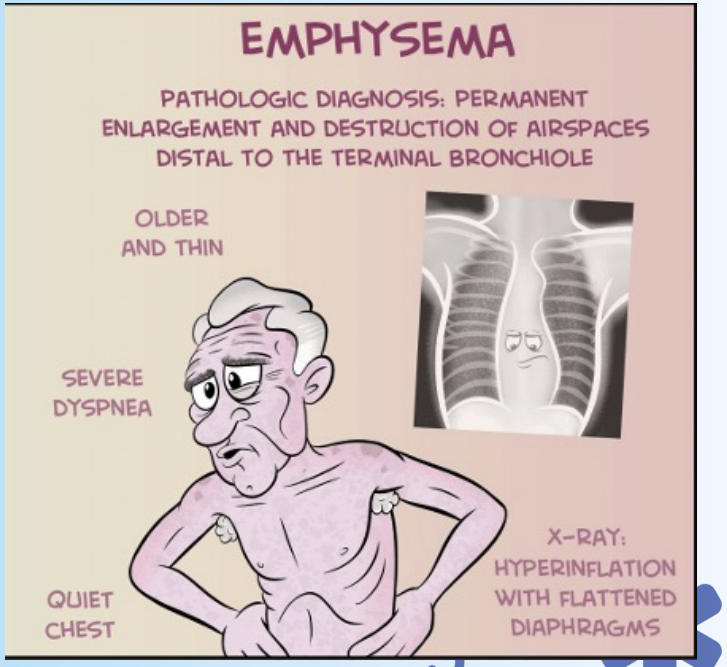
4. Airways collapse during ventilation

Causes “air trapping”

“The pink puffer”





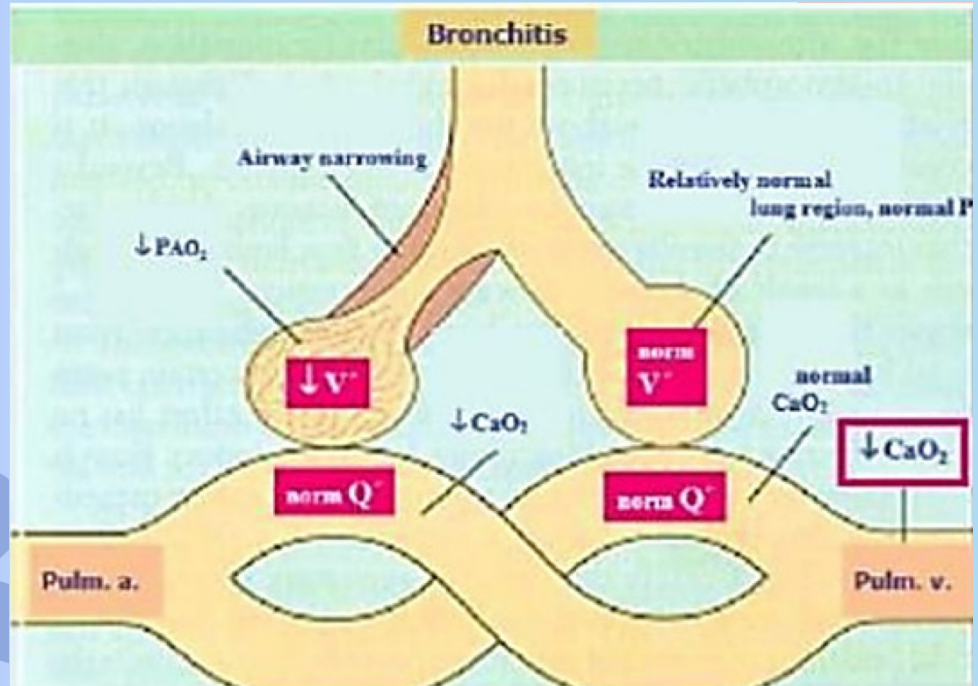


Summary

Chronic bronchitis
Cyanotic

Decreased alveolar O_2, but with normal perfusion causes decreased systemic O_2.

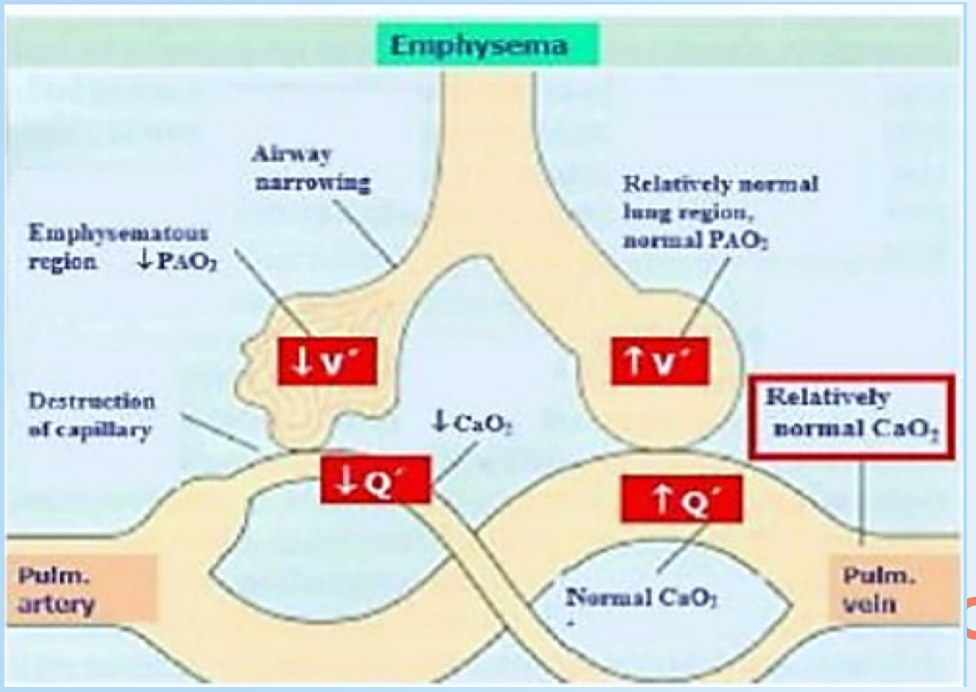
Decreased alveolar O_2 → ↑ Hemoglobin production



Emphysema
Non-cyanotic

Destruction of vessels causes a shunt towards functioning alveoli/ parts of the lungs.

Pulmonary hypertension → Backflow of blood into right ventricle → Right-sided heart failure



Chronic bronchitis
"The blue bloater"

Defined by clinical presentation:

Productive cough for at least 3 months over 2 consecutive years

Clinical signs and symptoms:

- Productive cough
- Cyanotic
- Wheezing/crackles on inspiration
- Overweight/peripheral edema
- Increased hemoglobin-levels
- Dyspnea

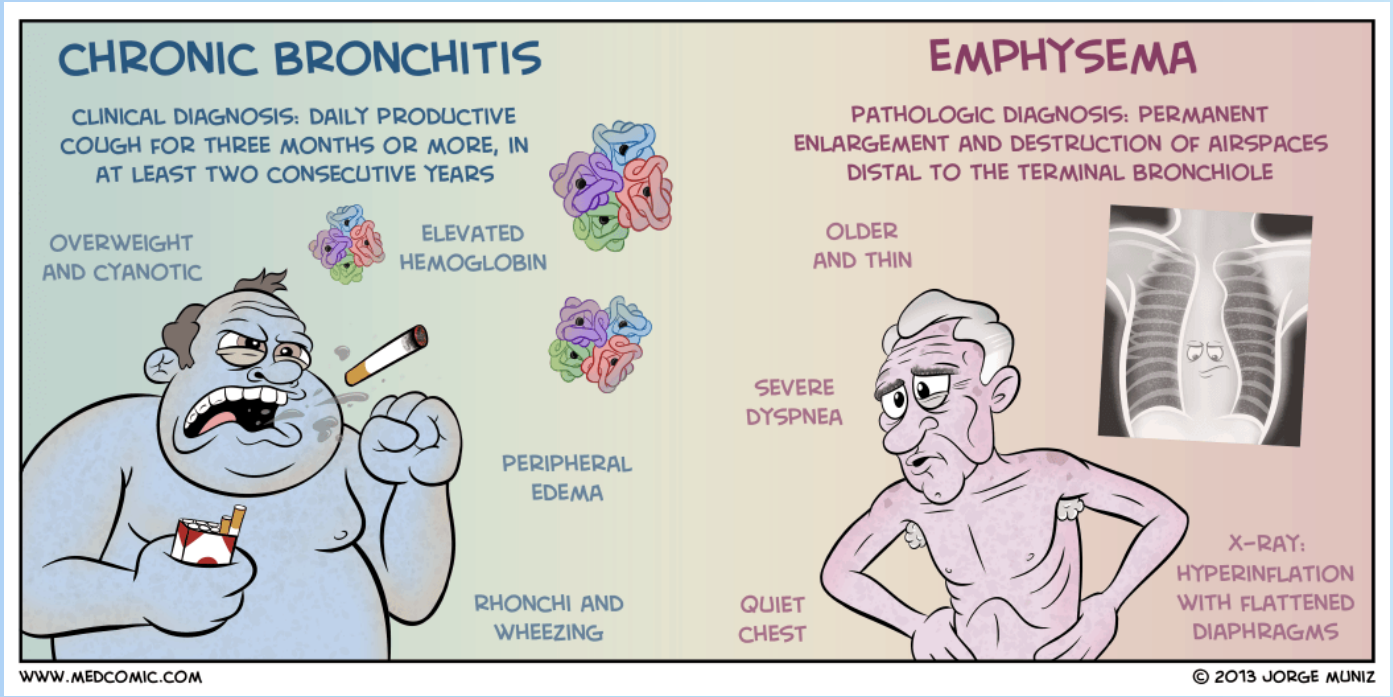
Emphysema
"The pink puffer"

Defined by structural presentation:

Permanent enlargement and destruction of pulmonary airways distally to the terminal bronchioles

Clinical signs and symptoms:

- Dyspnea
- Acyanotic, but pink skin
- Huge "Barrel chest"
- Skinny
- Pursed lip breathing

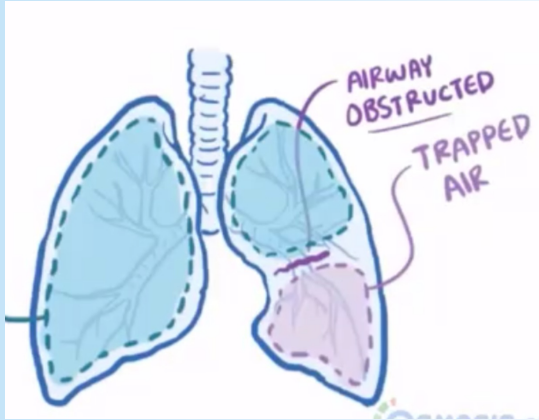


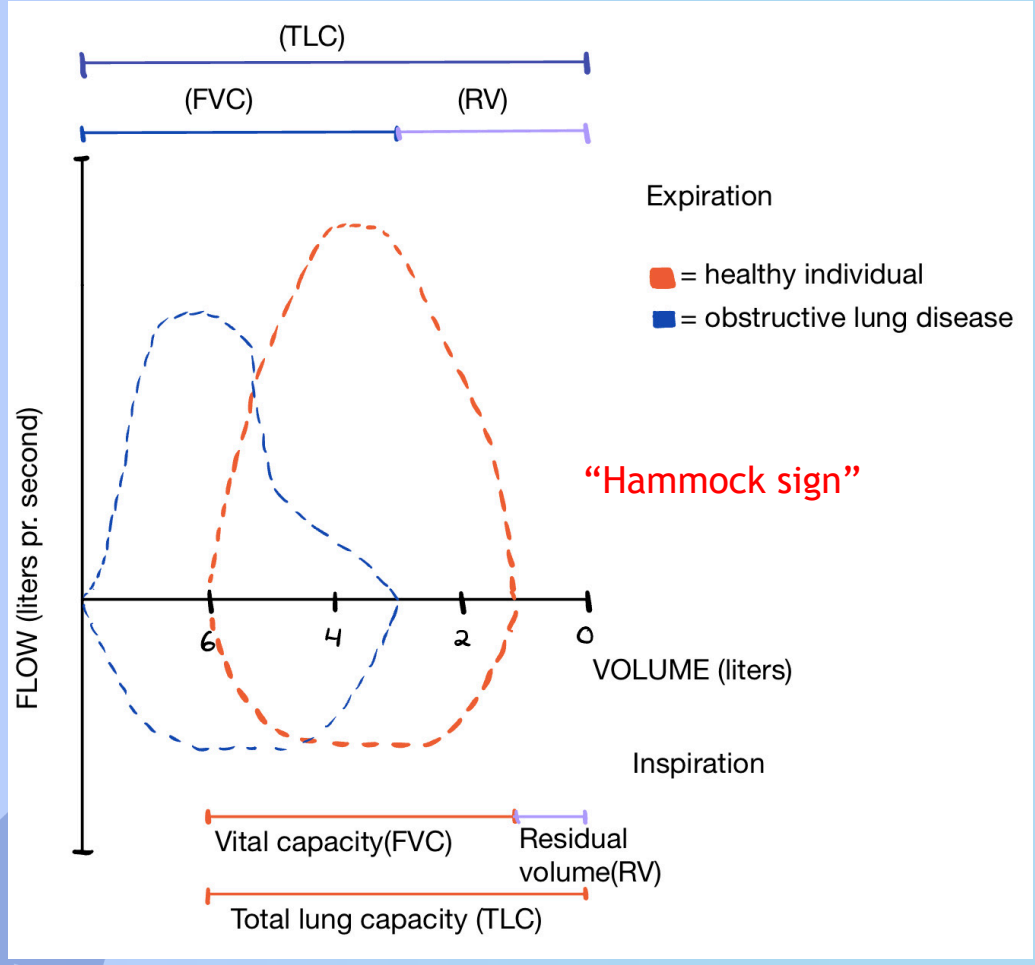
Pulmonary function test - characteristics

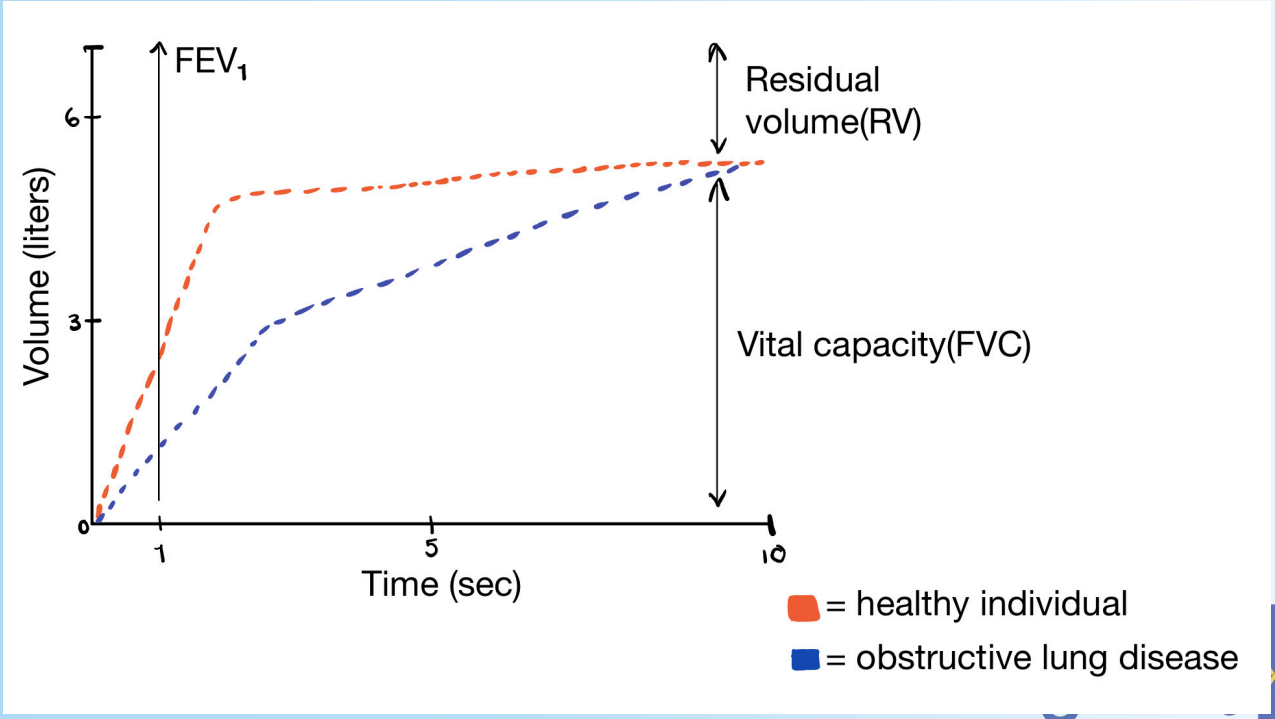
↑ Total lung capacity

↑ Residual volume

$$\frac{FEV1}{FVC} = <70\%$$







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~~→ Principe of V/Q~~

~~→ Lung volumes~~

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~~→ Asthma~~

~~→ COPD~~

3. Restrictive lung disorders

→ Idiopathic pulmonary fibrosis

→ Exposure-related restrictive lung diseases

→ Sarcoidosis

10 minute break

Restrictive lung disorders

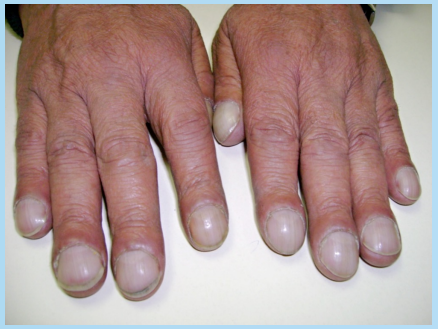
→ Idiopathic pulmonary fibrosis
→ Exposure-related restrictive lung disorders
→ Sarcoidosis

Idiopathic pulmonary fibrosis

Symptoms:

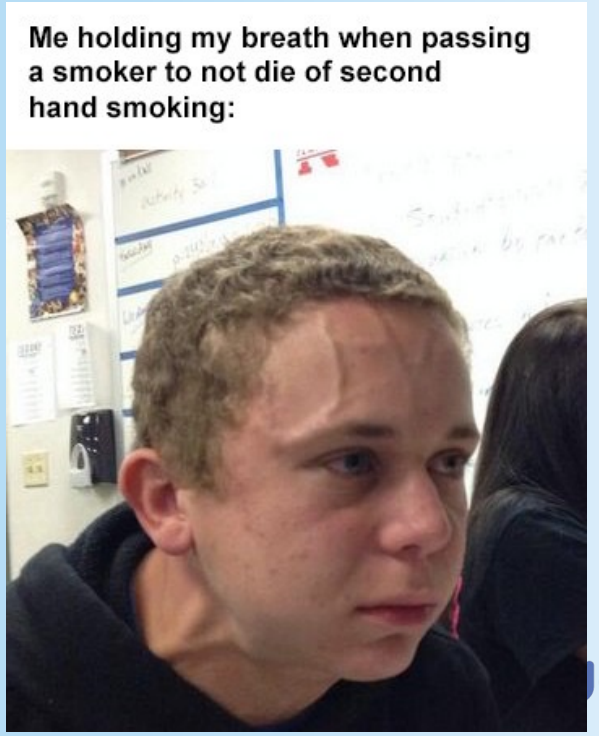
Progressive disorder – symptoms worsen over time

- Coughing
- Shortness of breath
- Cyanosis
- Digital clubbing
- Acid reflux/GERD
- Over time: Significant respiratory failure – lungs loose functional tissue



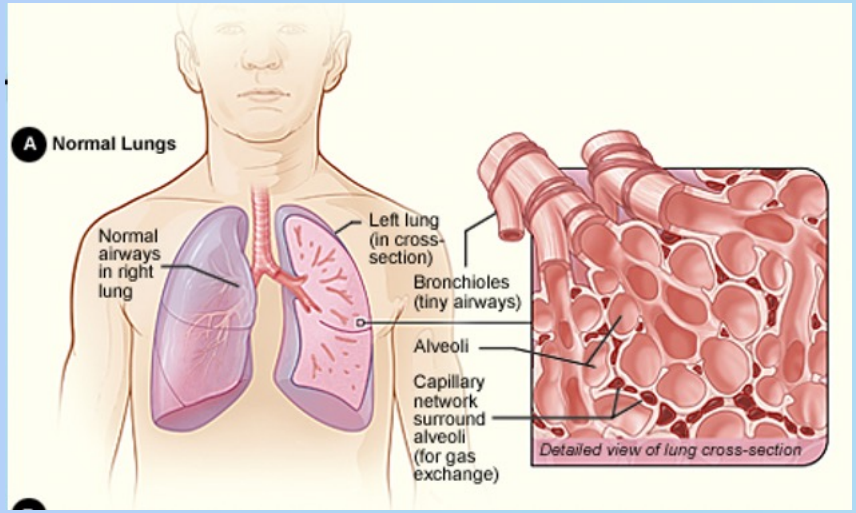
Risk factors:

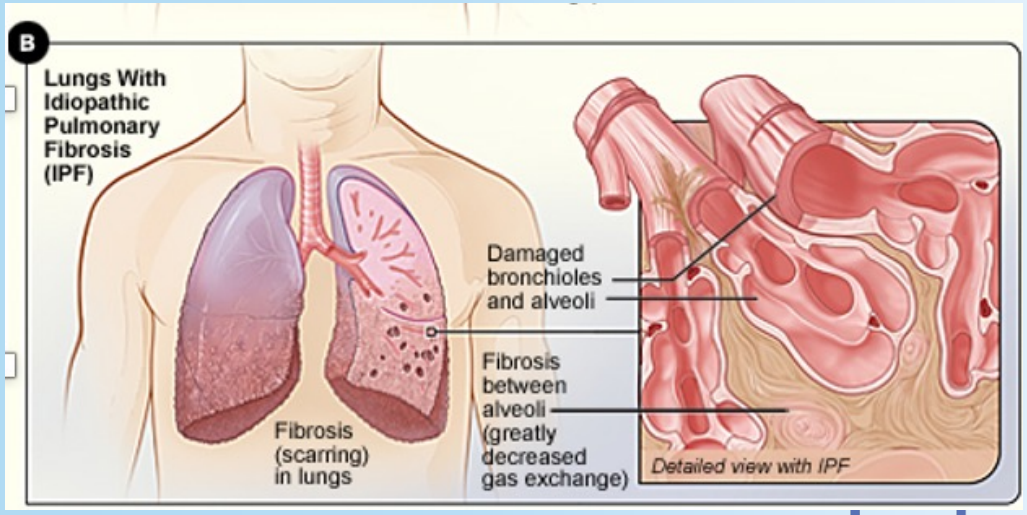
- Being male
- Old age
- Cigarette smoking



Pathogenesis of IPF

1. Damage to alveolar membrane
   Type 1-cells release TGFß-1
2. Type 1-cells stimulate type 2-cells
   Type 2 cells starts to proliferate uncontrollably
   Type 2 cells stimulate production of fibroblast
3. Uncontrollable production of fibroblast
   Fibroblast produce reticular fibers and elastic fibers
4. Thickening of interstitial layer
   Too much reticular fibers and elastic fibers lead to “stiff” lung, with no compliance





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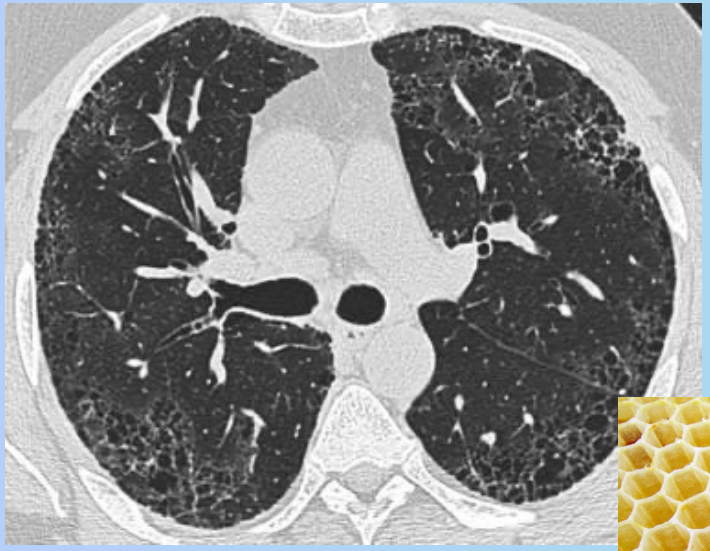
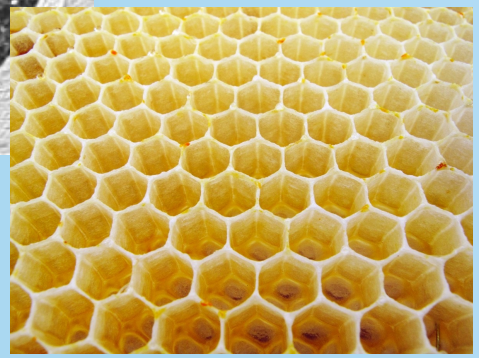
Diagnosis and treatment

- CT-scan
  - “Honeycomb appearance”
  - Thickening of interstitial walls





- Supplemental oxygen
- Antifibrotic medication
- Treat symptoms

  Acid reflux: Proton-pump inhibitors
  Hydrokodon: Cough
- Lung transplant

→ Only slows progression, does not stop the disease

Hypersensitivity pneumonitis

- Farmers lung
  Inhalation of actinomyces in hay
- Pigeon breeders lung
  Inhalation of proteins in bird poop or feathers
- Chemical worker`s lung

Symptoms:

Acute- type:

- Fever
- Shortness of breath
- Chest tightness
- Headaches

Chronic- type:

- Sustained shortness of breath

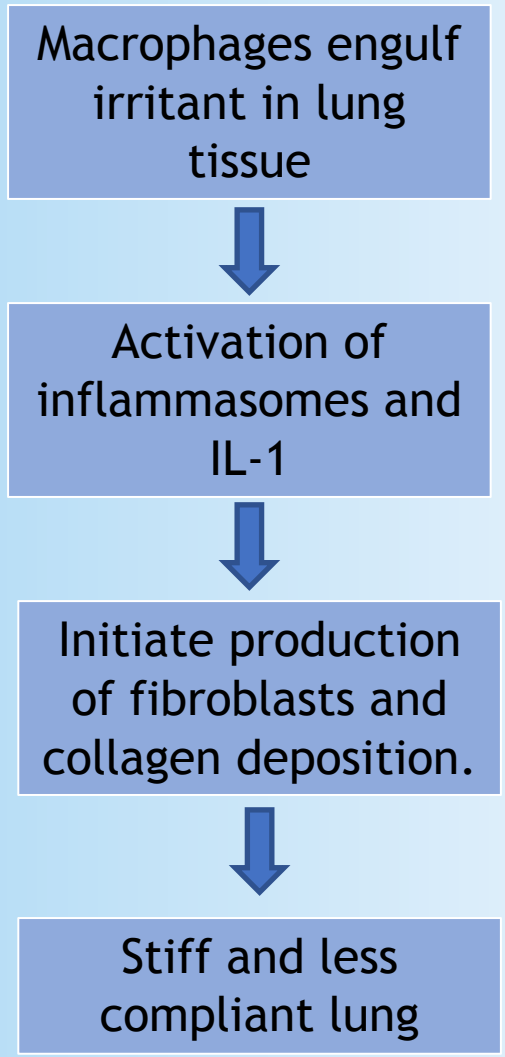
REVERSIBLE IF CAUGHT EARLY!

Hypersensitivity reaction → More fibrin, and less elastin Granulomatous inflammation → Stiff and less compliant lung

Pneumoconiosis

Exposure related restrictive lung disorder

- Asbestosis
  Caused by inhalation of asbestos
- Silicosis
  Caused by inhaling silica dust
- Coal worker`s pneumoconiosis
  Referred to as “black lung”
  Caused by inhaling coal mine dust

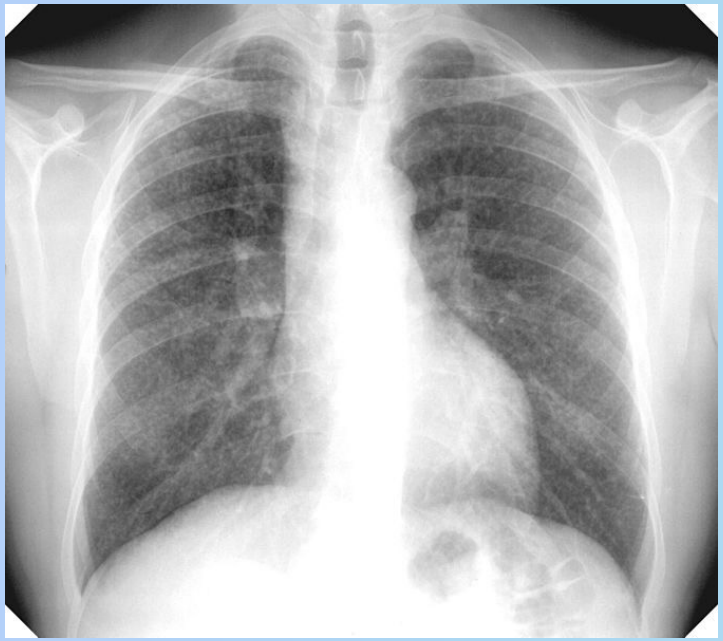


Diagnosis

(More important for pathomorphology)

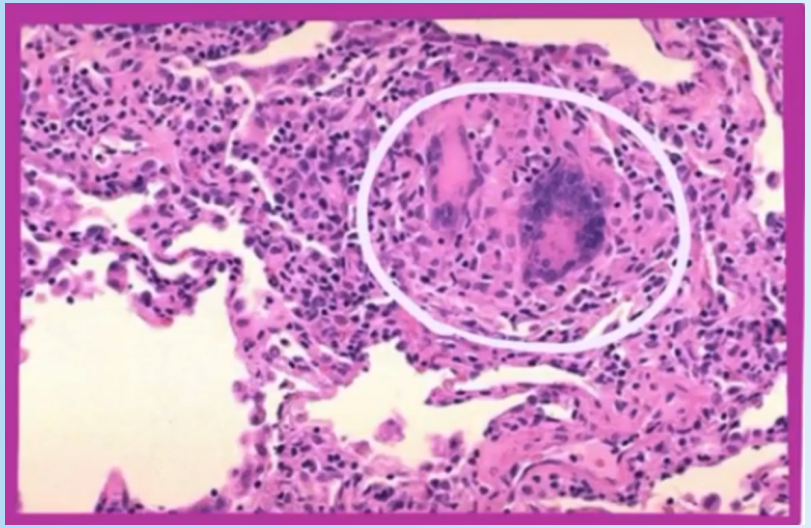
Chest X-ray

- Diffuse infiltrate



Biopsy of lung tissue (Bronchoalveolar lavage)

- Small granulomas
- Lymphocyte infiltration in alveolar walls



A day at your GP-office

70-year-old Mrs. Pani Biedronka comes in to your GP-office for her routine checkup. She is not a woman that likes to complain, but today she has a concern she wants to share with you. Lately she has been experiencing loss of breath when she goes to the bathroom, fever and coughing. She has a family history of restrictive lung disease and had tuberculosis and Lyme disease as a child.

AUSCULTATION
- Bilateral crackles heard at base of lung

BLOOD TEST
↑ Calcium & ↑ ACE

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Sarcoidosis

Your stereotypical patient:

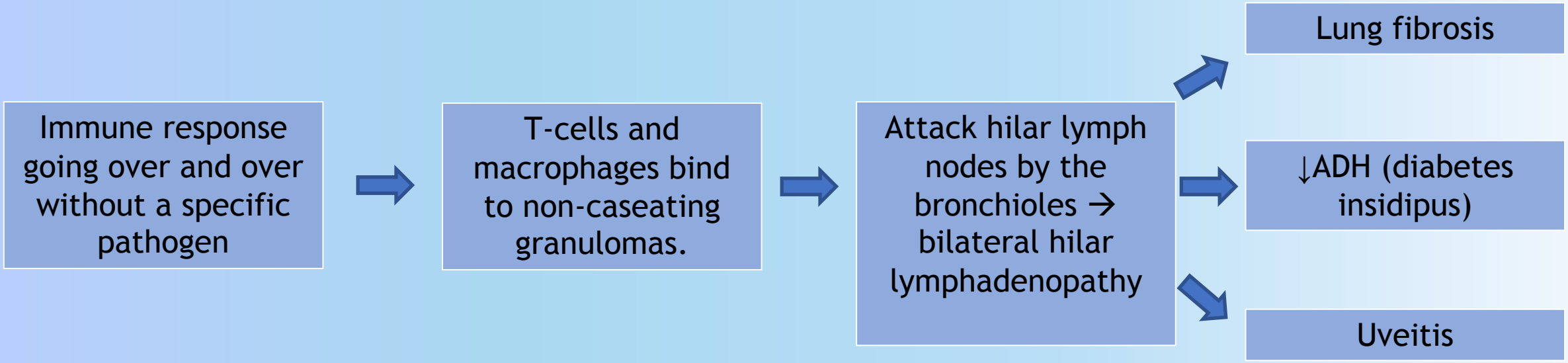
- African woman
- Patient with family history of sarcoidosis
- Patient that has previously been exposed to tuberculosis or Borrelia (lyme disease)

Symptoms:

Usually appear asymptomatic

- Fever
- Weight loss
- Fatigue
- Shortness of breath & coughing
- Tender leg nodules
- Vision changes

Pathogenesis of sarcoidosis



- Pathogenesis is not fully understood.
- Systemic disease characterized by non-ceasing granulomas
- The non-caseating can attack everywhere in the body.

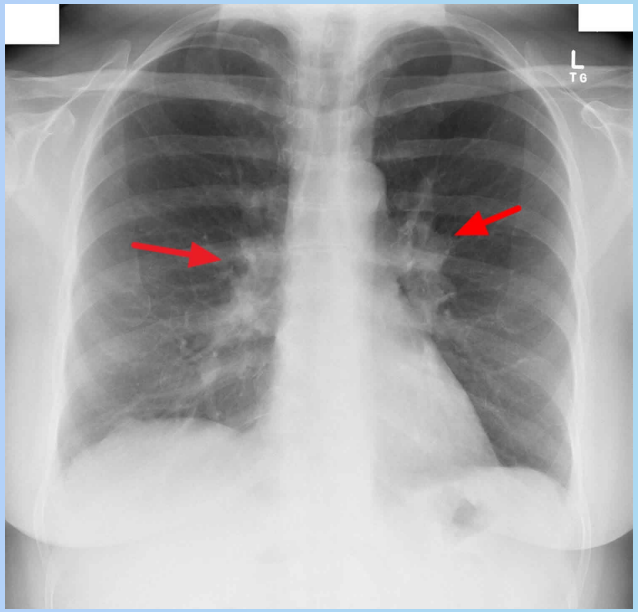
Pathomorphology note:
The non-caseating granulomas contain:
- No necrotic tissue
- Fibroblasts
- Lymphocytes
- Fibrous tissue
- Multinucleated giant cells
- Schaumann`s bodies
- Asteroid bodies

Diagnosis

(more important for pathomorphology)

Chest X-ray or CT-scan

- Bilateral hilar lymphadenopathy



Blood tests

↑ Calcium

Excess vitamin D from macrophages

↑ Angiotensin converting enzyme (ACE)

Produced by T-cells

Bronchoalveolar lavage (biopsy)

↑ T-cells infiltration in lung

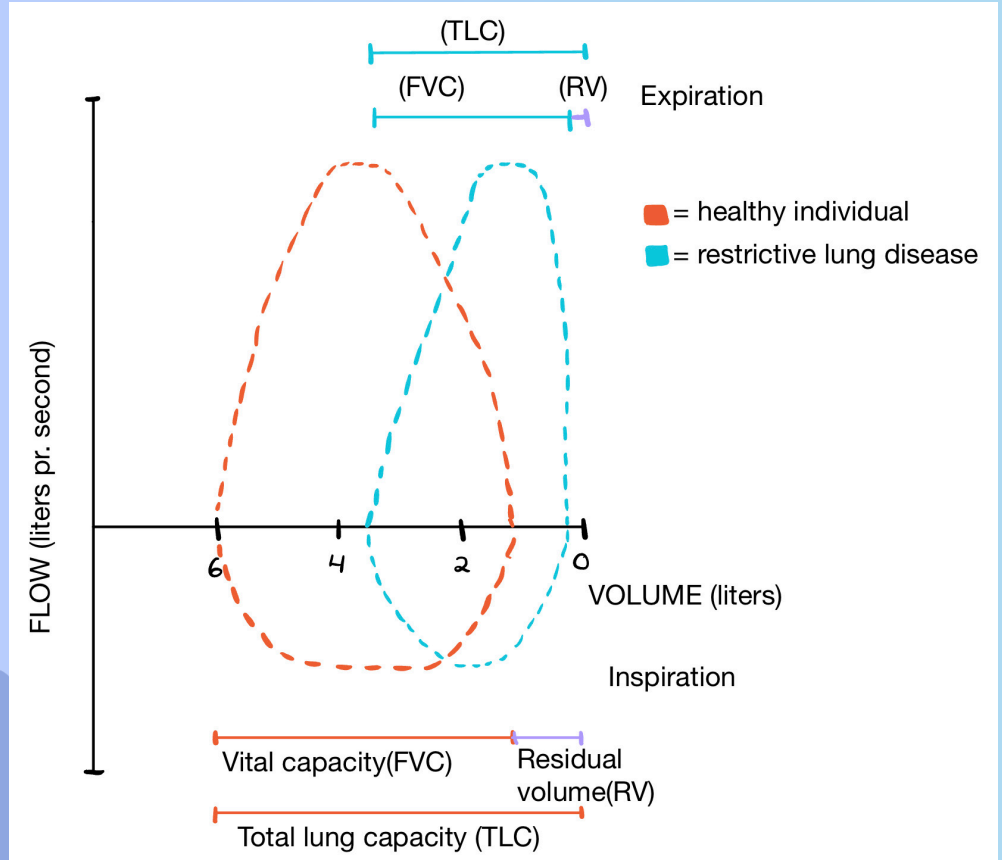
Pulmonary function test- characteristics

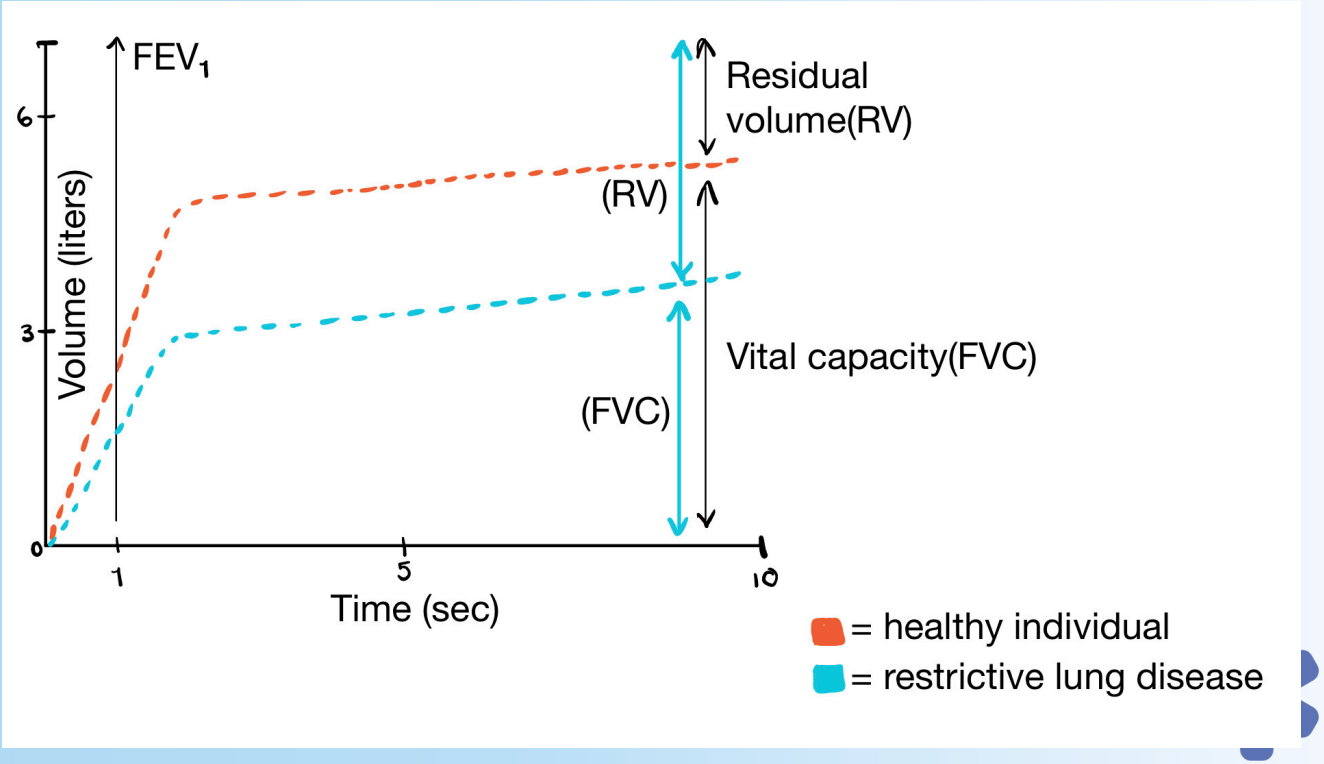
↓ Total lung capacity

↓ Forced vital capacity

↓ Residual volume

$\frac{FEV1}{FVC}$ = Normal/slightly decreased





	Obstructive Lung diseases		Restrictive Lung diseases	
	Asthma	COPD (Emphysema + Chronic Bronchitis)	Idiopathic Pulmonary Fibrosis	Sarcoidosis
Clinical Presentation	Young child, difficulty breathing (dyspnea), audible wheezing (on expiration) and chest tightness	Dyspnea with history of smoking, barrel shaped chest Productive cough	Older patient with progressive dyspnea and a dry cough	asymptomatic young woman.
Symptoms + Pathogenesis	Dyspnea + chest tightness – Bronchoconstriction from Bronchospasm (due to mast cell degranulation) Wheezing – mucus plugs that obstruct exhalation	Dyspnea – Collapse of bronchioles due to loss of elastic fibers (Emphysema) Productive cough – Mucus gland hyperplasia and hypersecretion	Progressive dyspnea – Excessive fibrosis cause decreased lung compliance	Non-caseating granulomas in different parts of the body may cause symptoms
Diagnosis	Clinical symptoms that are reversible on administration of Bronchodilators (β2-agonists)	Emphysema – X-Ray Chronic Bronchitis is clinical – Greater than 3 months of productive cough in 2 consecutive years	Exclusion "honeycomb appearance"	Exclusion
Spirometry	FEV1 = ↓↓ FVC = ↓ or normal FEV1/FVC = ↓	FEV1 = ↓↓ FVC = ↓ or normal FEV1/FVC = ↓	FEV1 = ↓ or Normal FVC = ↓↓ FEV1/FVC = Normal or ↑	FEV1 = ↓ or Normal FVC = ↓↓ FEV1/FVC = Normal or ↑