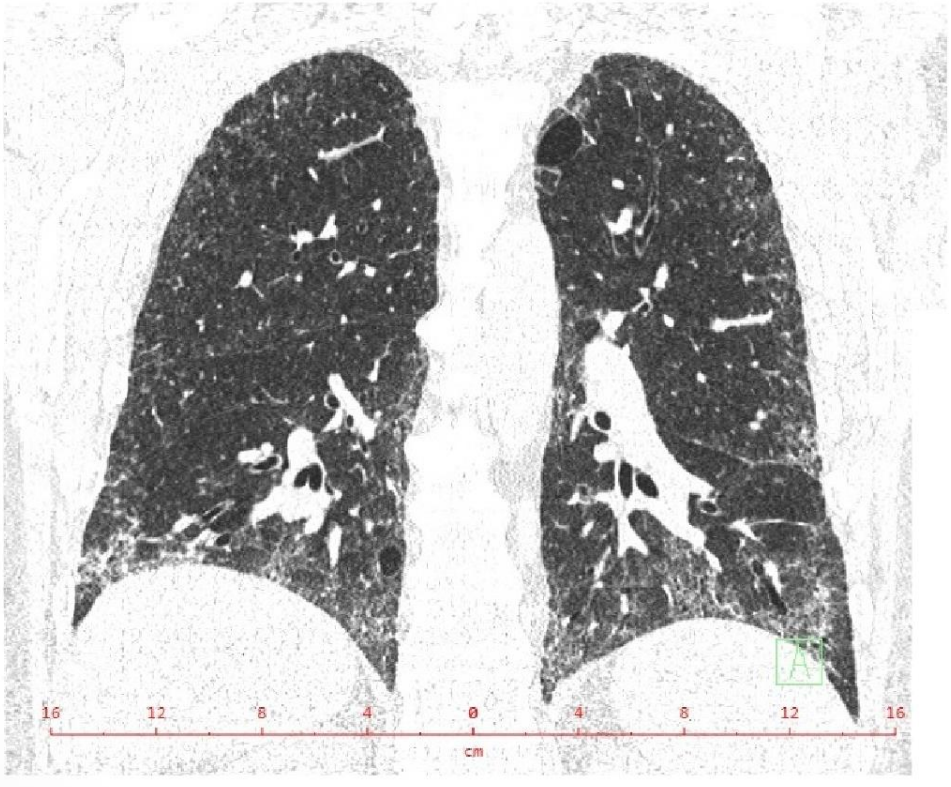
Pathology of the lungs 3

MUDr. Jan Balko, Ph.D.

Department of Pathology and Molecular Medicine,
2nd Faculty of Medicine, Charles University in Prague and
Motol University Hospital



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Lung malformations

Lung malformations

- prenatal anatomical abnormality of the lungs, exceeding the level of variability

bronchial stenosis / atresia

bronchogenic cyst

pulmonary hypoplasia

CPAM / CCAM

pulmonary sequestration

Bronchial stenosis / atresia

Definition

- stenosis to atresia of the bronchial lumen

Causes (etiology)

- congenital defect of luminisation (partial / complete)

Developement (pathogenesis)

- variable respiratory restriction based on the locality (peripheral can be asymptomatic; central significant)

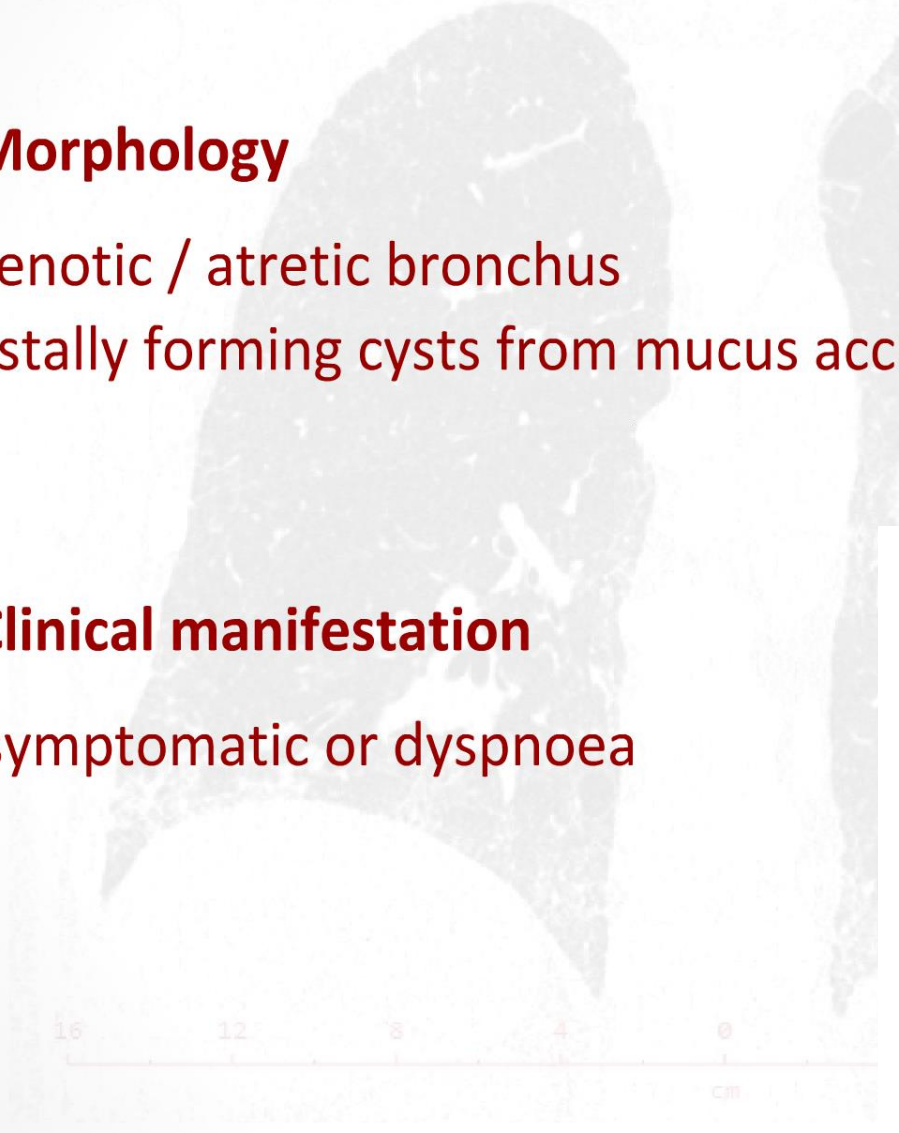
Bronchial stenosis / atresia

Morphology

- stenotic / atretic bronchus
- distally forming cysts from mucus accumulation

Clinical manifestation

- asymptomatic or dyspnoea



Bronchogenic cyst

Definition

- cyst covered with bronchial mucosa

Causes (etiology)

- (extra)pulmonal detachment of embryonal pulmonary cells

Developement (pathogenesis)

- cysts are formed in the locus of detachment (analogy to the bronchial wall)

Bronchogenic cyst

Morphology

- mediastinal / pulmonary (detached from bronchial tree)
- cyst filled with mucus (several cm in diameter)
- **microscopically** bronchial wall (smooth muscle, cartilage...)

Clinical manifestation

- asymptomatic or compression of the thoracic organs

Pulmonary hypoplasia

Definition

- congenitally small lungs (most common pulm. malformation)

Causes (etiology)

- compression (oligohydramnios, diaphragmatic hernia) / trizomy (12; 18; 21) / chest deformity (bone dysplasia)

Developement (pathogenesis)

- low respiratory surface leading to the RI

Pulmonary hypoplasia

Morphology

- small lungs (weight comparsion)
- lower lobes do not overhang heart

Clinical manifestation

- asymptomatic or hydrops and fetal death

CPAM / CCAM

Definition

- CPAM = congenital pulmonary airway malformation
- CCAM = congenital cystic adenomatoid malformation

Causes (etiology)

- congenital disorder of the bronchioalveolar units formation

Developement (pathogenesis)

- mucus accumulation within bronchioalveolar units disconnected from bronchial tree and their **cystic dilatation**

CPAM / CCAM

Morphology

- children (rarely adults)
- focal / diffuse cystic dilatation of the lung parenchyma
- **microscopically** 6 types of the cysts (size, epithelium...)

Clinical manifestation

- recurrent infection and dyspnoea
- rarely mucinous metaplasia and even adenocarcinoma of the lung in childhood

Pulmonary sequestration

Definition

- a piece of pulmonary parenchyma not attached to the lung

Causes (etiology)

- isolation of embryonal pulmonary cells with independent developement

Developemant (pathogenesis)

- bronchiectasis and mucostasis are developed in time, unable to cough up the mucus

Pulmonary sequestration

Morphology

- intra- / extrapulmonal p. parenchyma separated from the lung
- blood supply from the aberrant artery from aorta
- not communicating with bronchial tree

Clinical manifestation

- recurrent infection
- volume overload of the small circulation because of abnormal blood supply (pulmonary hypertension)

Vascular lung diseases

Vascular lung diseases

- a group of diseases characterised with disorder of **pulmonary circulation** (small blood circulation)
- the majority can be acute or chronic (except DAH)

pulmonary edema
pulmonary embolism (+ lung infarction)
pulmonary hypertension
DAH
pulmonary vasculitis

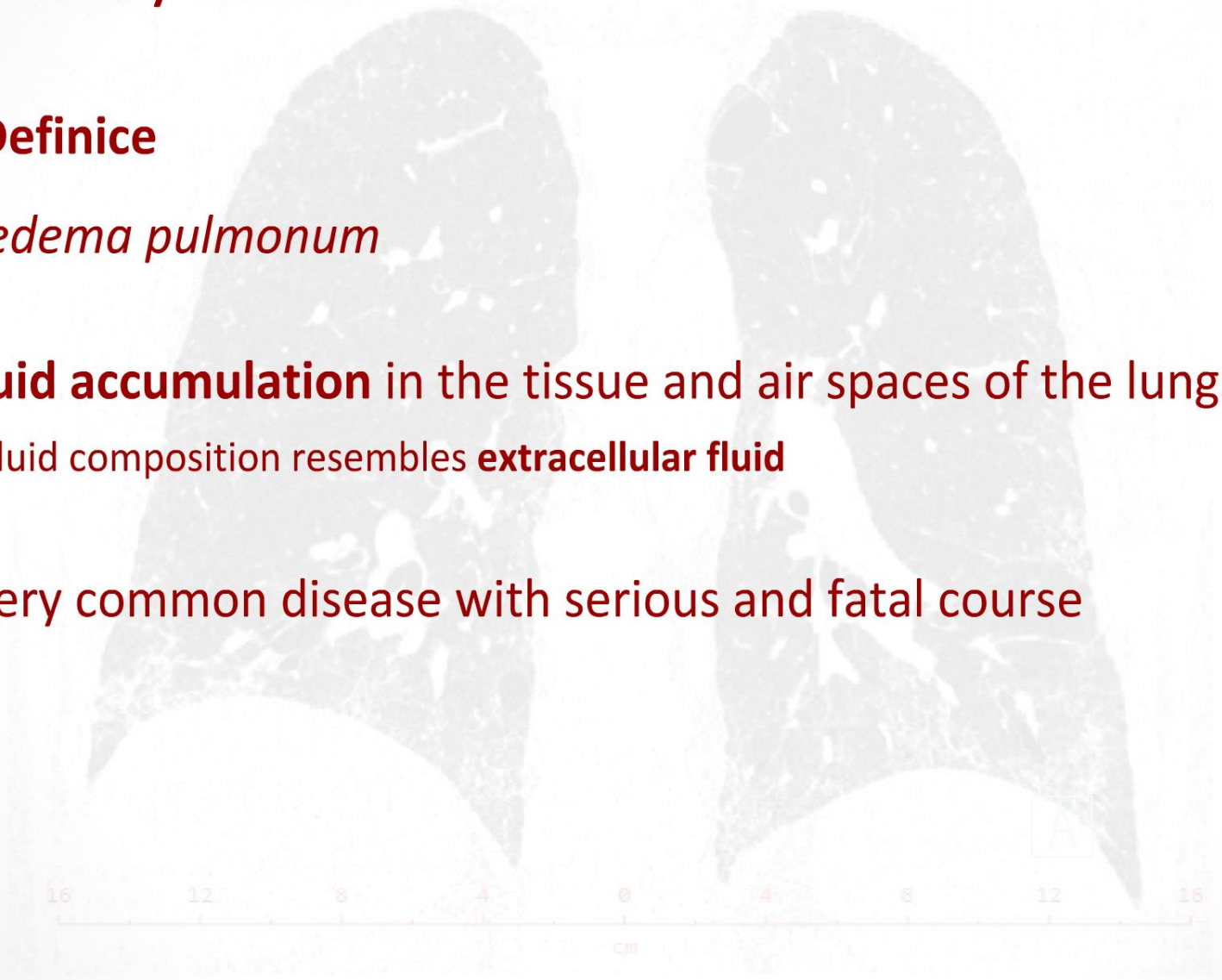
Pulmonary edema

Definice

- *oedema pulmonum*

- **fluid accumulation** in the tissue and air spaces of the lungs
  - fluid composition resembles **extracellular fluid**

- very common disease with serious and fatal course



Pulmonary edema

Causes (etiology)

- **hemodynamic** = due to the disruption of Starling's forces
  - ↑ **hydrostatic pressure** = left heart failure (the most common one)
  - ↓ **oncotic pressure** = burns, nephrotic syndrome, uremia
  - **neurogenic** = disruption of the autonomic pathways within the spinal cord and capillary dilatation (CNS defects)

- **cytotoxic** = caused by damage of capillary wall
  - **endogenous** = shock, inflammation (even autoimmune)
  - **exogenous** = inhalation (toxic substances, hot air), lung trauma

- the combination of causes is most commonly observed

Pulmonary edema

Developement (pathogenesis)

- presence of fluid leads to **impaired gas exchange** and changes in **pressure rates**
  - pulmonary hypertension catalyzes hypertrophy of tunica media in **vessels**
  - **interstitium** undergoes fibrosis, which intensfies pulmonary hypertension
  - hypertension translates to the right **heart** (*cor translatum*)

- **complications** can develope
  - **brown induration** = fibrosis and siderophage deposition within the lungs due to the long standing pulmonary congestion
  - **hypostatic bronchopneumonia** = infection of edematous fluid
  - **carnifications** = organisation of fibrin in edematous fluid

Pulmonary edema

Morphology

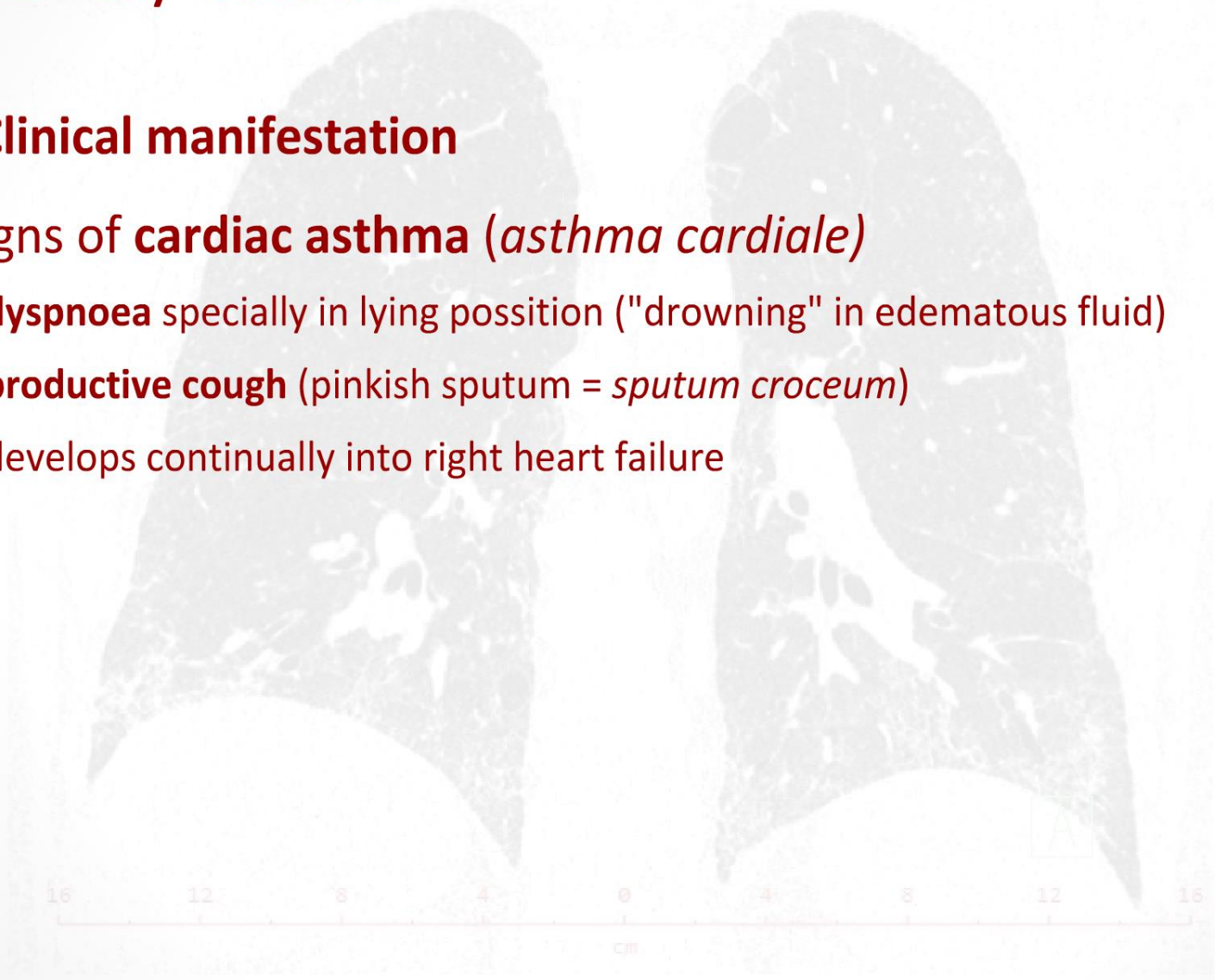
- **macroscopically** heavy lung soaked with fluid
  - LL over 480 g and RL over 570 g (sometimes even more than 1 kg)
  - lungs are filled with **foamy fluid**

- **microscopically** alveoli contain granular eosinophilic fluid
  - **acute** = plasma escapes in alveolar spaces + congested capillaries
  - **chronic** = rupture of capillaries leads to release of hemosiderin from damaged erythrocytes (engulfed by siderophages) + induration
  - cytotoxic edema can contain inflammatory cells and capillary microthrombi

Pulmonary edema

Clinical manifestation

- signs of **cardiac asthma** (*asthma cardiale)*
  - **dyspnoea** specially in lying possition ("drowning" in edematous fluid)
  - **productive cough** (pinkish sputum = *sputum croceum*)
  - develops continually into right heart failure



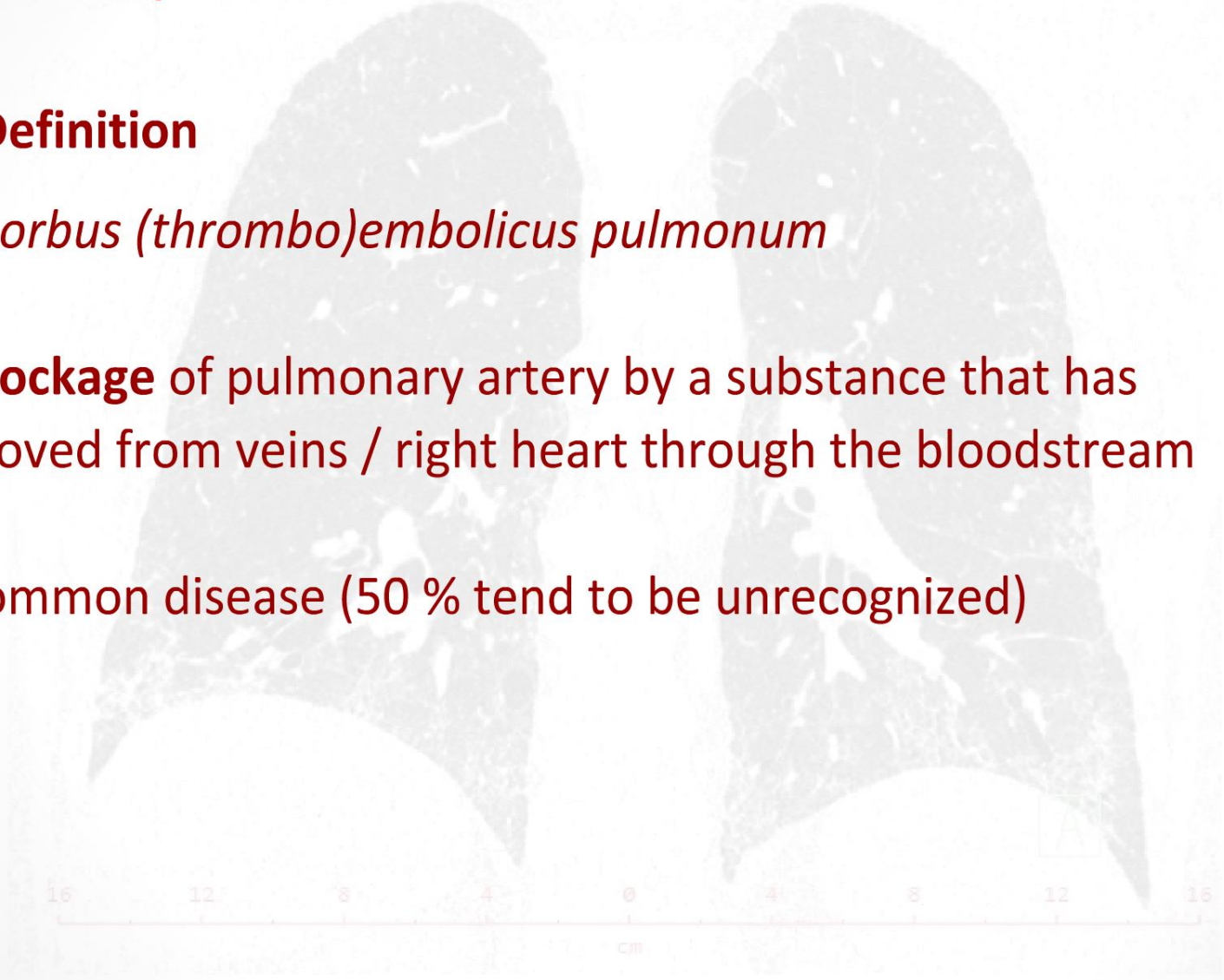
Pulmonary embolism

Definition

- *morbus (thrombo)embolicus pulmonum*

- **blockage** of pulmonary artery by a substance that has moved from veins / right heart through the bloodstream

- common disease (50 % tend to be unrecognized)



Pulmonary embolism

Causes (etiology)

- various **materials** can embolize:
  - **thrombi** (= thromboembolism, blood clot *intra vitam*, mainly veins of LE)
  - **infected trombi** (peripheral pyemia as a complication of TF)
  - **air** (trauma, operation with aspiration over 10 ml of air)
  - **azote** (caisson disease / decompression sickness)
  - **fat** (trauma of long bones, polytrauma of subcutis, burns)
  - **bone marrow** (trauma of axial bones)
  - **amniotic fluid** (complicated delivery, anaphylaxis to fetal Ags)
  - **heterogenous material** (catheter, talc in cases of drug-addicts)
  - **tumour cells** (angioinvasion)

Pulmonary embolism

Developement (pathogenesis)

- determined according to the **size of embolus**:

- **massive** = a. pulmonalis and its branches
  - increases blood pressure in the right heart with obvious dilatation of RA + RV and developement of **cor pulmonale acutum**)
  - usually sudden death

Pulmonary embolism

Developement (pathogenesis)

- determined according to the **size of embolus**:

- **submassive** = smaller branches of a. pulmonalis
  - lower increase of right heart pressure with dilatation of RA + RV and developement of **cor pulmonale acutum**)
  - right heart failure is also possible, but slower

- can lead to the **pulmonary infaction**
  - only under condition of heart failure (insufficiency of nutritive and functional pulmonary circulation)
  - sphenic **hemorrhagic necrosis** and fibrinous pleuritis (followed by scarring)
  - **complication** secondary anaerobic infection (pulmonary gangrene)

Pulmonary embolism

Developement (pathogenesis)

- determined according to the **size of embolus**:

- **successive** = capillaries (chronic, repeatedly)
  - gradual pulmonary hypertension without dilatation of RA + RV, but with hypertrophy = **cor pulmonale chronicum**)
  - heat failure (slow) and pulmonary infarction (numerous) are also possible

Pulmonary embolism

Morphology

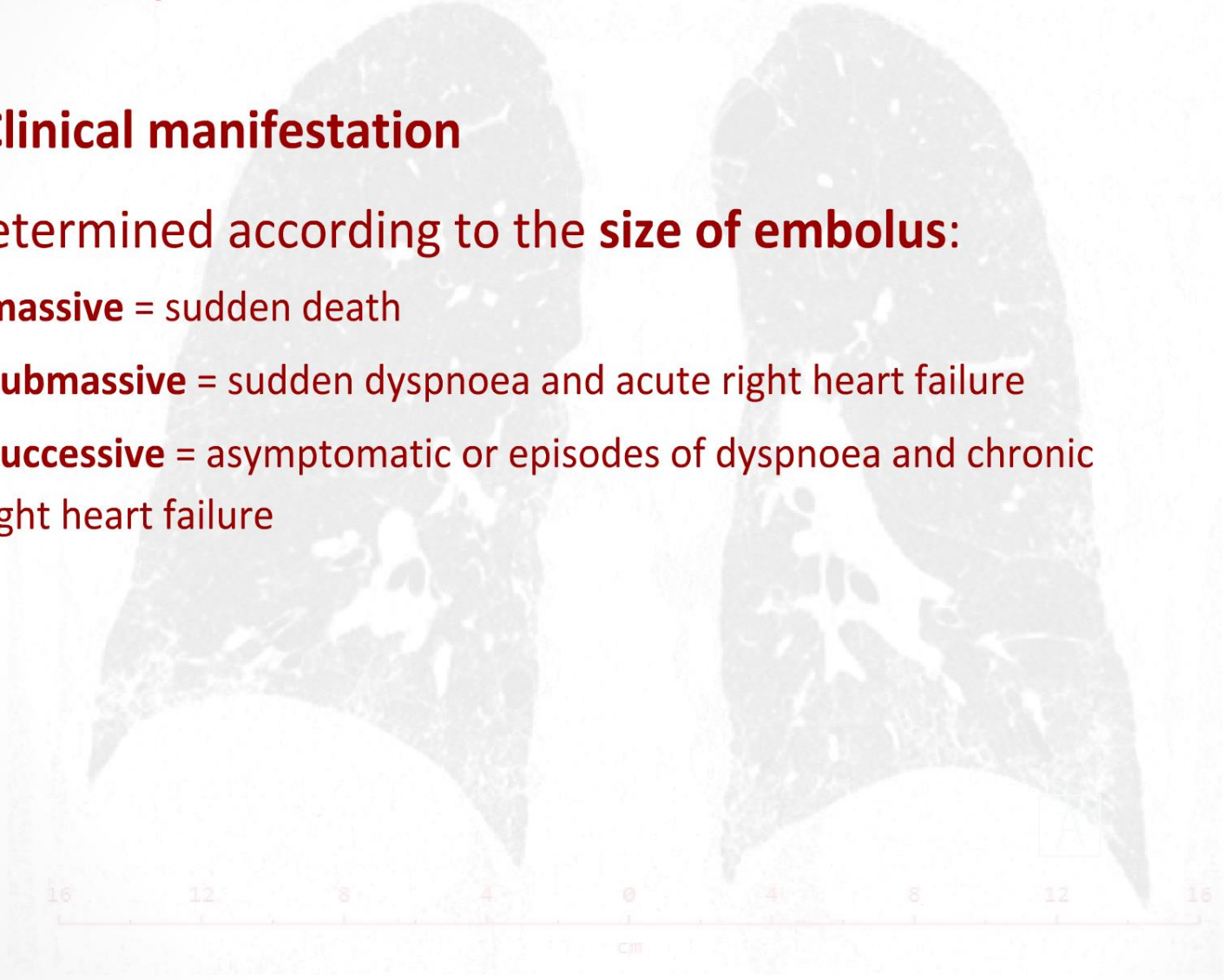
- **macroscopically** obturation of p. arteries with trombus
  - compared to **cruor**, the thrombus is solid, fragile and brashy + connected to the vessel wall
  - **pulmonary infarction** looks sphenic and red
  - **chronic** one shows hallmarks of healing

- **microscopically** obturation of p. arteries with trombus
  - organisation or even recanalization of larger thrombi (vital reaction)
  - **pulmonary infarction** represents hemorrhagic necrosis
  - the proof of **fat** embolism requires frozen cut

Pulmonary embolism

Clinical manifestation

- determined according to the **size of embolus**:
  - **massive** = sudden death
  - **submassive** = sudden dyspnoea and acute right heart failure
  - **successive** = asymptomatic or episodes of dyspnoea and chronic right heart failure



Pulmonary hypertension

Definition

- *morbus hypertonicus pulmonum*

- increased blood pressure within the pulmonary artery **over 30 mmHg**

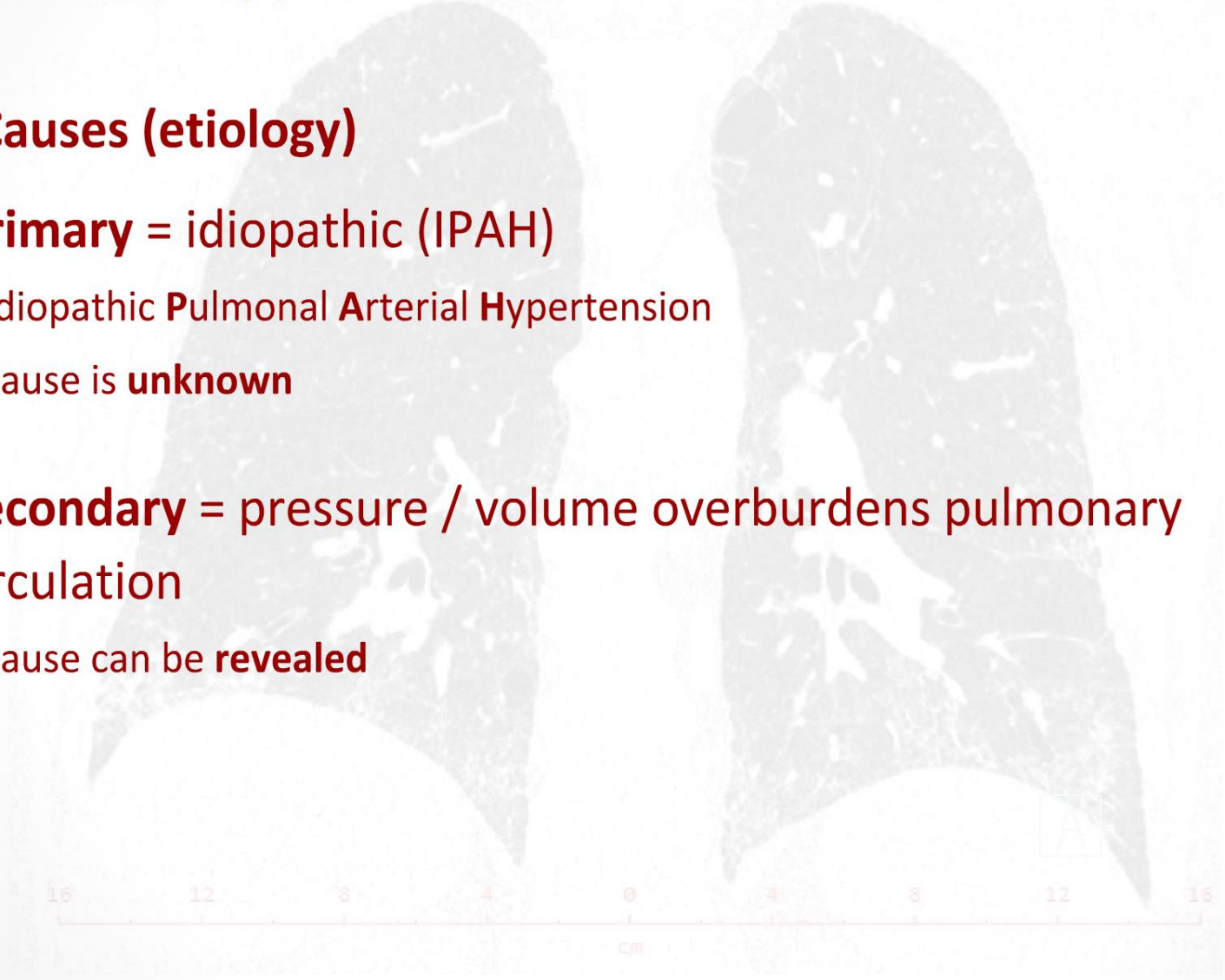
- primary (rare, but fatal), secondary (common)

Pulmonary hypertension

Causes (etiology)

- **primary** = idiopathic (IPAH)
  - **I**diopathic **P**ulmonal **A**rterial **H**ypertension
  - cause is **unknown**

- **secondary** = pressure / volume overburdens pulmonary circulation
  - cause can be **revealed**



Pulmonary hypertension

Developement (pathogenesis)

- **precapillary** = disorder enters via aa. pulmonales
  - IPAH, cardiac left-to-right shunts (higher V of blood in RV), pulmonary embolism, IDLs

- **postcapillary** = disorder enters via vv. pulmonales
  - cor translatum, pulmonary veno-occlusive disease

- allways leads to the overloading of RV and **cor pulmonale chronicum**

Pulmonary hypertension

Morphology

- **macrscopically** resembles fibrous atherosclerotic plaques
  - visible in **large pulmonary arteries**

- **micrscopically** remodeling of **small pulmonary vessels**
  - hypetrophy of tunica media of arterioles → proliferation of tunica intima of arterioles (stenosis) → lumen obturation → plexiform arteriopathy → dilatation or bleeding → fibrinoid necrosis

Pulmonary hypertension

Clinical manifestation

- **primary** = young adult ♀
  - **incurable** with gradual progression towards right heart failure

- **secondary** = older adults
  - **curable** with gradual progression towards right heart failure

DAH

Definition

- diffuse alveolar hemorrhage

- **acute massive bleeding** into lung interstitium and alveoli
  - must be bilateral and developed at the same time (otherwise there is a different source of bleeding)
  - part of **acute lung injury (ALI)** next to the ARDS

- rare, but fatal

DAH

Causes (etiology)

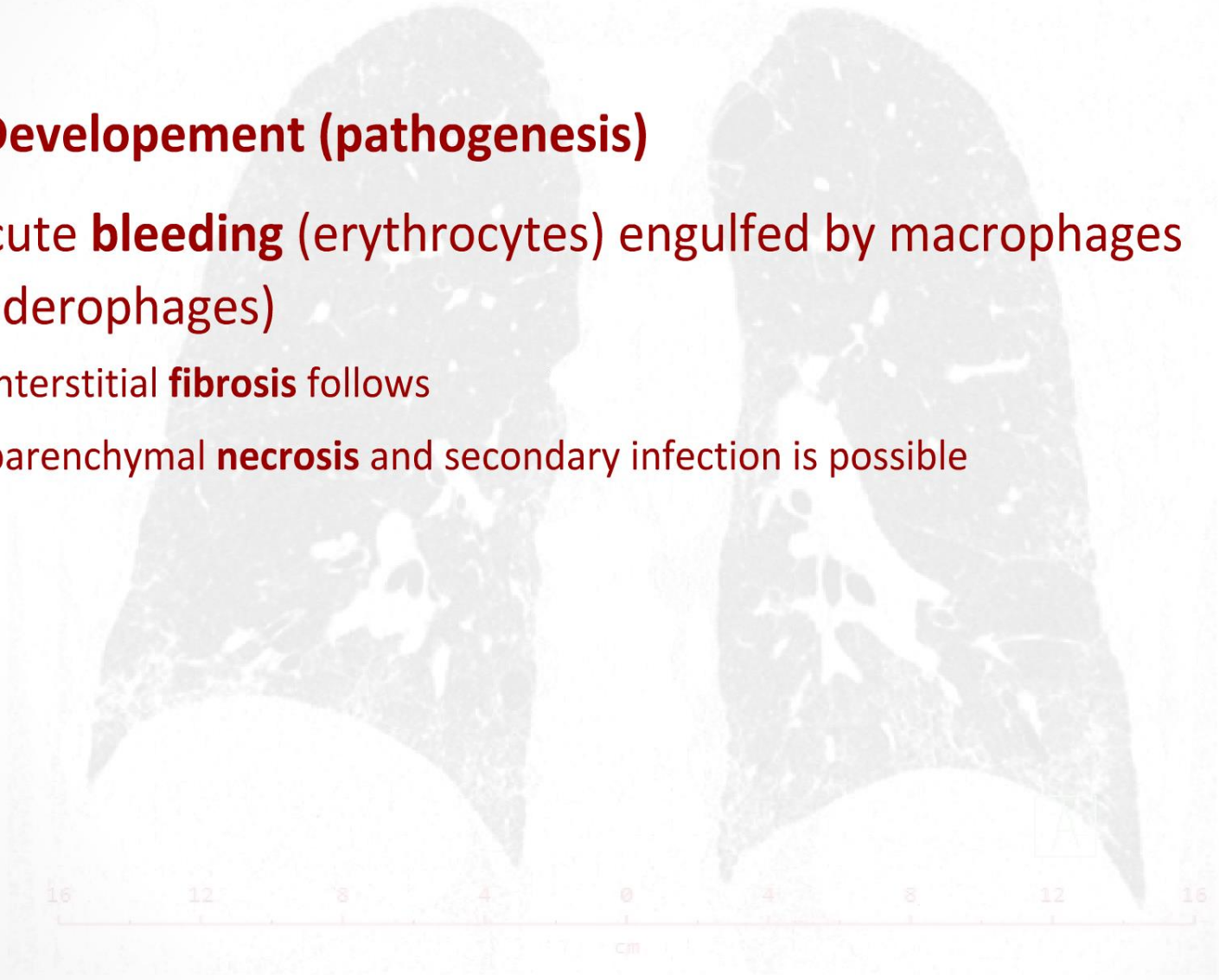
- **immune** causes = usually autoimmune mechanism
  - **Goodpasture syndrome**, collagenosis, vasculitis

- **unknown** cuases = idiopathic
  - so called idiopathic alveolar hemorrhage / isolated pulmonary pauciimmune capillaritis

DAH

Developement (pathogenesis)

- acute **bleeding** (erythrocytes) engulfed by macrophages (siderophages)
  - interstitial **fibrosis** follows
  - parenchymal **necrosis** and secondary infection is possible

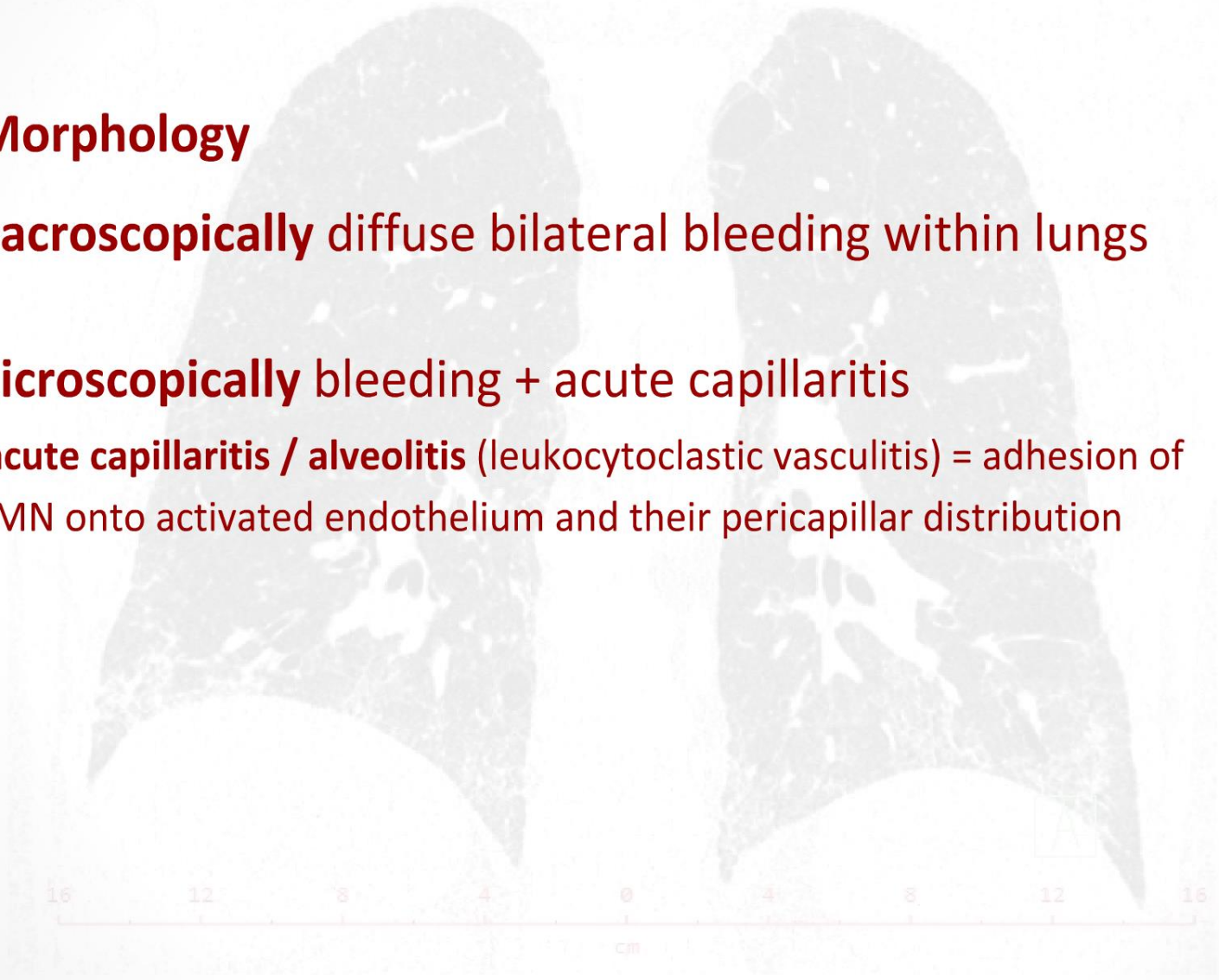


DAH

Morphology

- **macroscopically** diffuse bilateral bleeding within lungs

- **microscopically** bleeding + acute capillaritis
  - **acute capillaritis / alveolitis** (leukocytoclastic vasculitis) = adhesion of PMN onto activated endothelium and their pericapillar distribution



DAH

Clinical manifestation

- allways **severe condition**
  - dyspnoea with **hemoptysis** and life-threatening RI

Atelectasis / collapsed lung

Atelectasis / collapsed lung

Definition

- absence of the air within lung parenchyma

- **atelectasis** = focally collapsed lung
  (used to stand for collapsed lung of the newborns)

- **collapsed lung** = min. 1 lobe of the whole lung is collapsed
  (used to stand for collapsed lung which has already been providing gas exchange)

Atelectasis / collapsed lung

Causes (etiology)

- **atelectasis**:

1) obstruction

- closure of the ascendent brochus for a *part of the lung*
  - **intraluminal** (aspiration, tumor, mucus plug, blood clot)
  - **extraluminal** (tumor, lymph node, inflammation)

2) compression

- pressing a *part the lung* from the outside
  - **pathological pleural content** (hydro- / hemo- / pyothorax)

3) low surface tension (surfactant deficiency)

- allveoal collapse after 1st inbreath of immature newborns

Atelectasis / collapsed lung

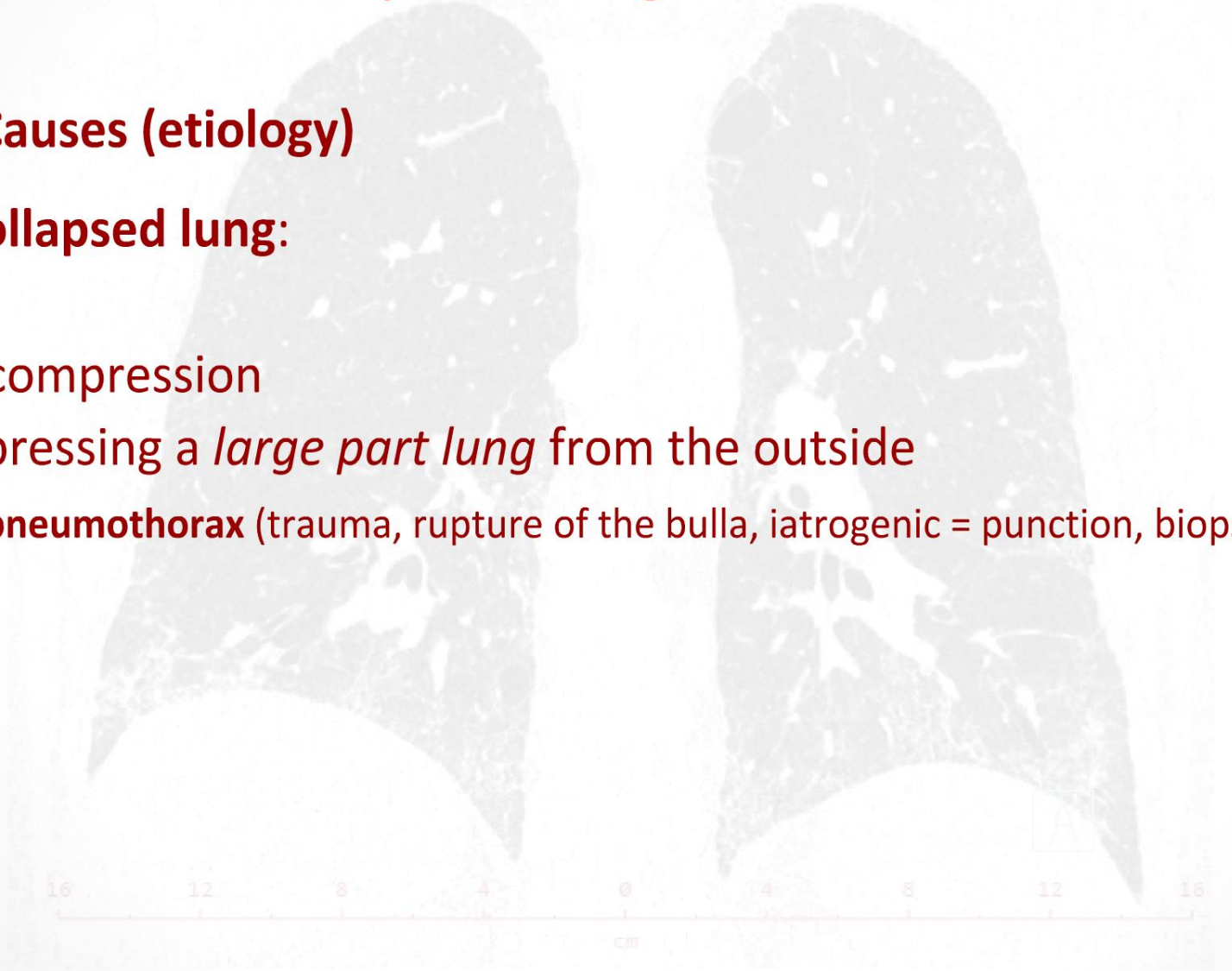
Causes (etiology)

- **collapsed lung**:

1) compression

- pressing a *large part lung* from the outside

• **pneumothorax** (trauma, rupture of the bulla, iatrogenic = punction, biopsy)



Atelectasis / collapsed lung

Developement (pathogenesis)

- air **resorption** and **alveolar collapse**
  - air gets absorbed into blood

Atelectasis / collapsed lung

Morohology

- **macroscopically** firm collapsed parenchyma without air
  - often lower lobes / surroundings of the rapidly growing lesions (pneumonia, tumor bleeding)
  - in time, **collapse induration** of the lung is developed ("splenisation")
  - high position of the diaphragm, wrinkled pleura, medistinal shift
  - water test in newborns (except from *pulmo spumosus*)

- **microscopically** collapsed alveoli
  - slit-like spaces with activated pneumocytes followed by fusion of their walls (induration)

Atelectasis / collapsed lung

Clinical manifestation

- depends on extension (atelectasis can be asymptomatic; collapsed lung leads to RI)
  - usually **dyspnoea, cyanosis, tachypnoe**
  - collapse can be converted into partial one (dilatation due to the adhesions) by talc aplication into pleural cavity

- **acute**
  - inflation of the parenchyma after elimination of the cause (reversibile)

- **chronic**
  - fibrotisation (induration) of the lung parenchyma (ireversibile)

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