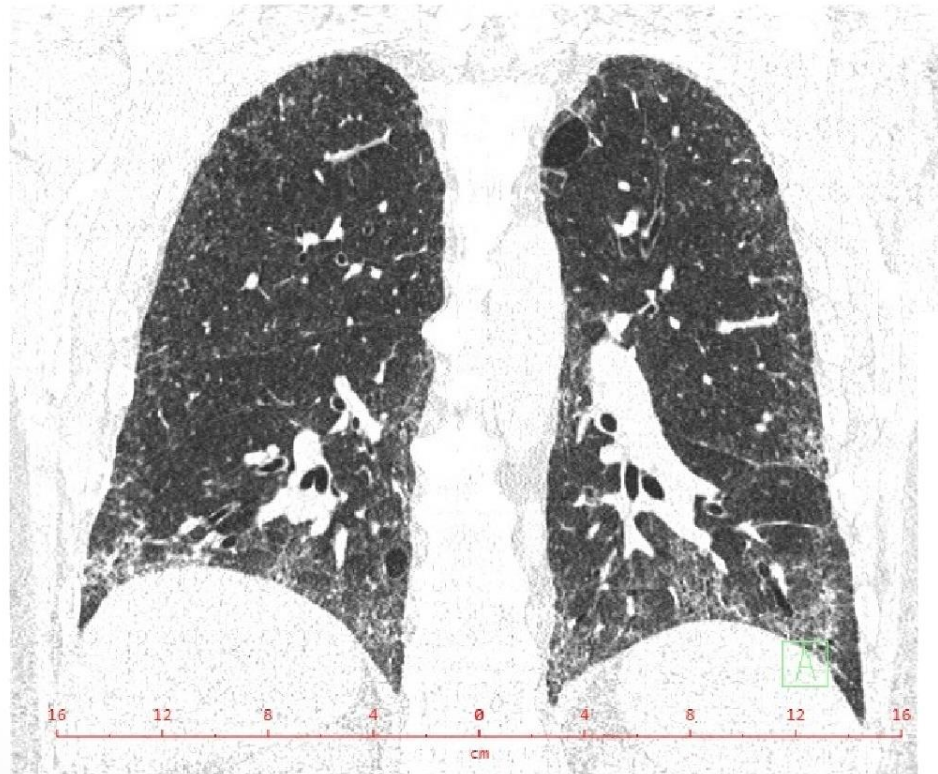


# Pathology of the lungs 3

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**FN MOTOL**



**2. LF UK**

# Diseases of the lungs

**1) malformations**

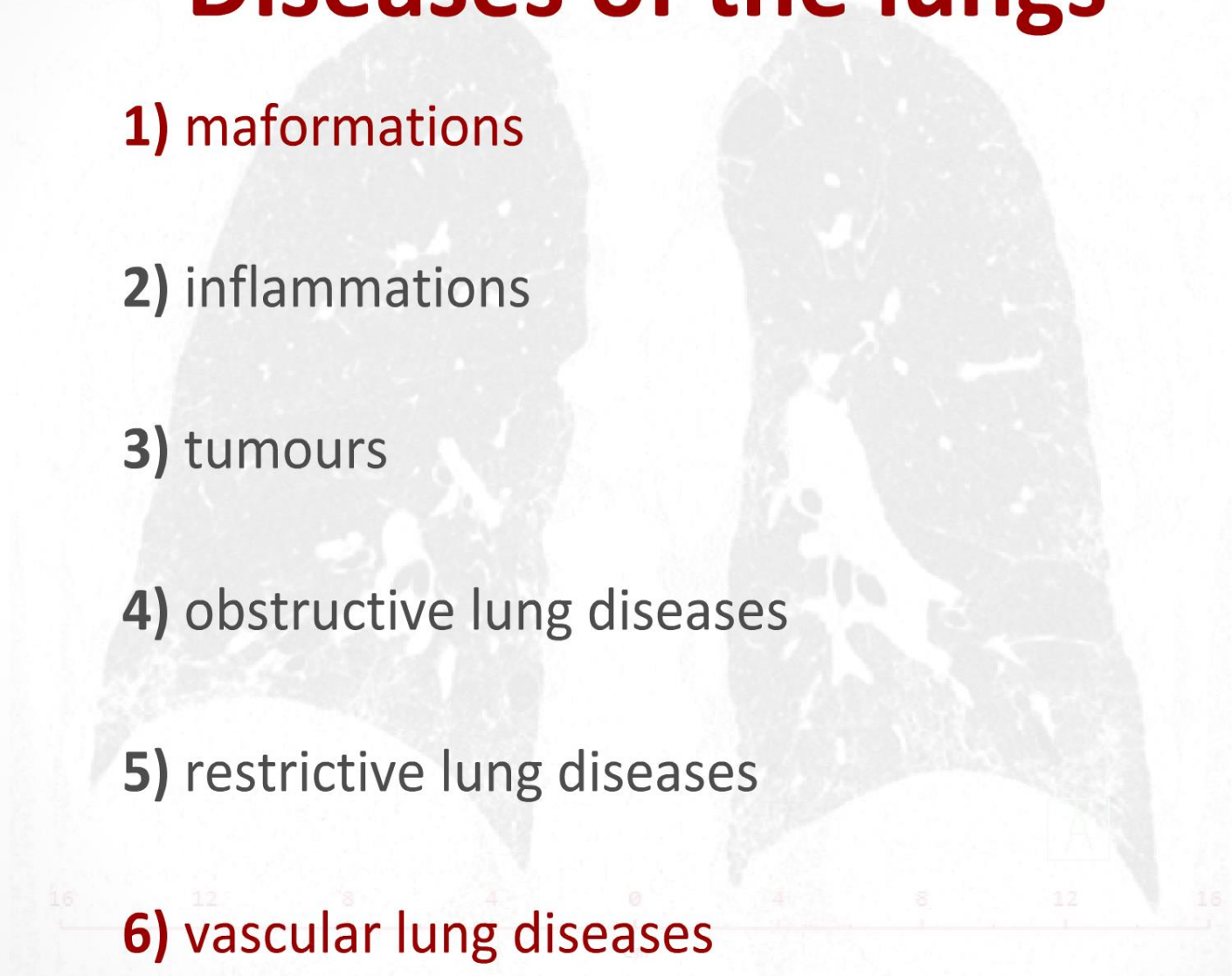
**2) inflammations**

**3) tumours**

**4) obstructive lung diseases**

**5) restrictive lung diseases**

**6) vascular lung diseases**



# Lung malformations



# Lung malformations

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

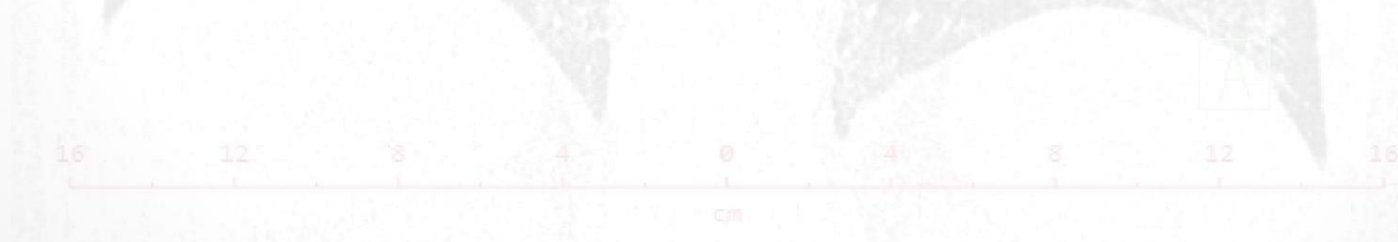
bronchial stenosis / atresia

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)
- mediastinal / pulmonary (detached from bronchial tree)

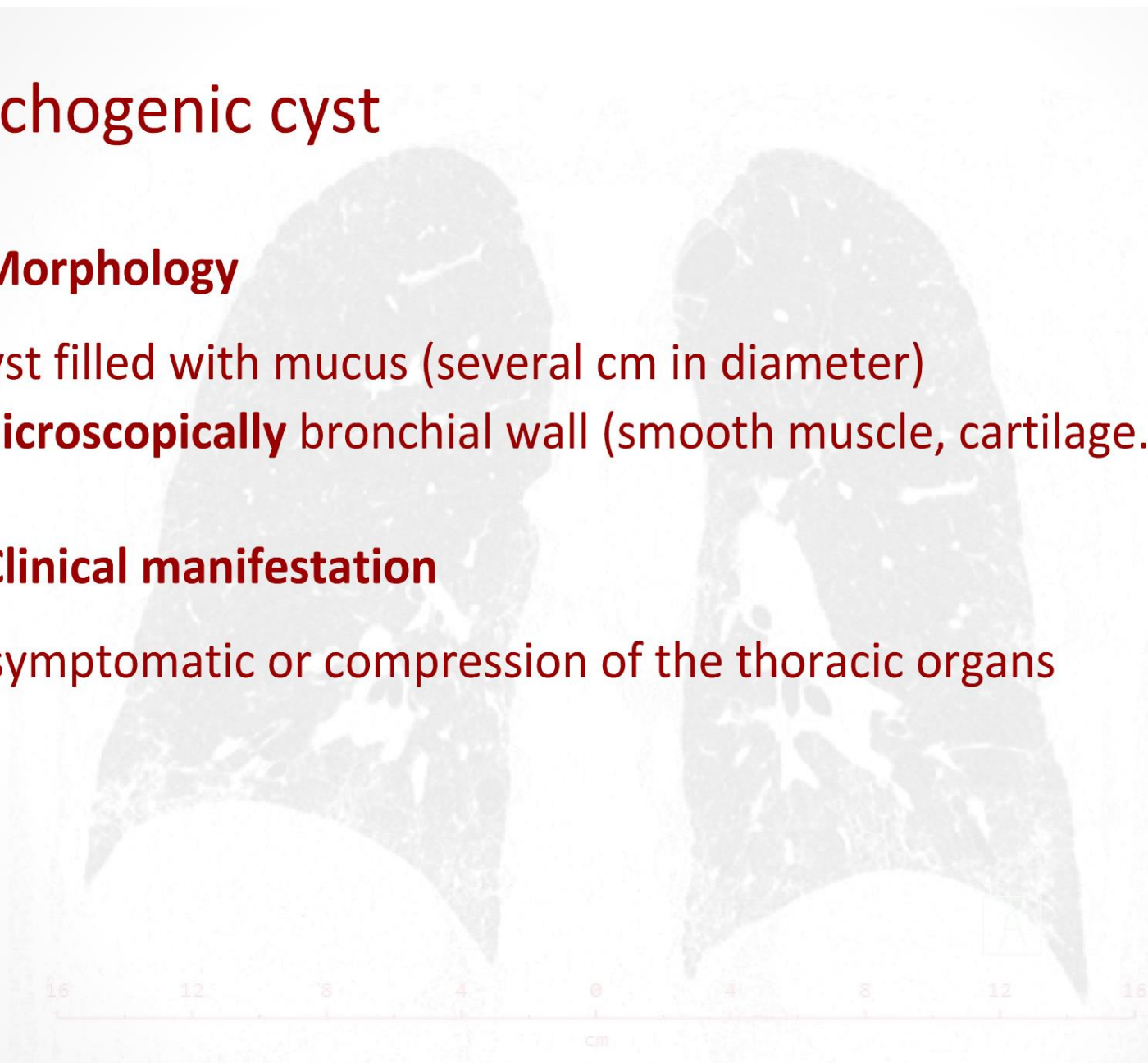
# Bronchogenic cyst

## Morphology

- 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

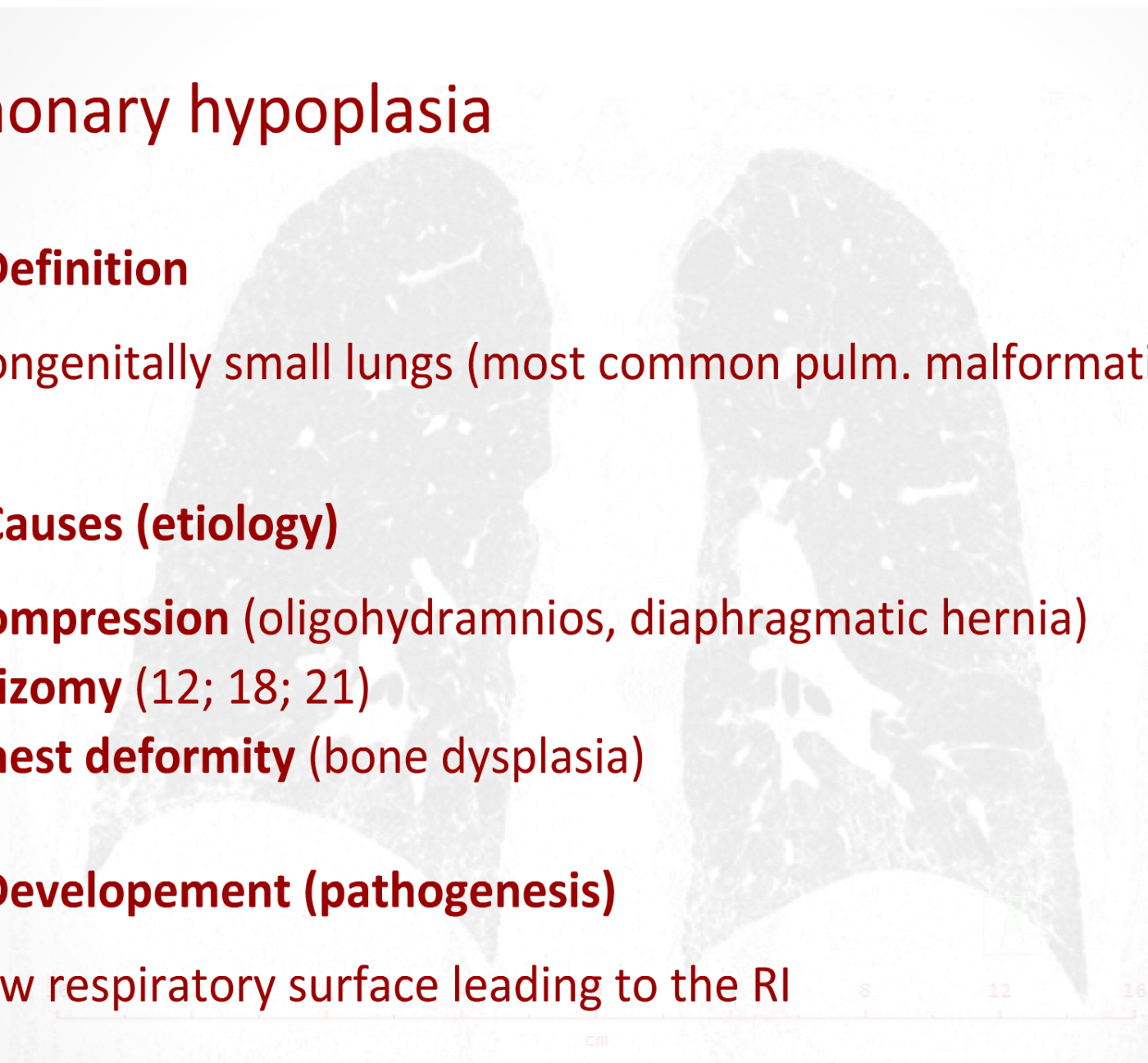
- congenitally small lungs (most common pulm. malformation)

## 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



## Development (pathogenesis)

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

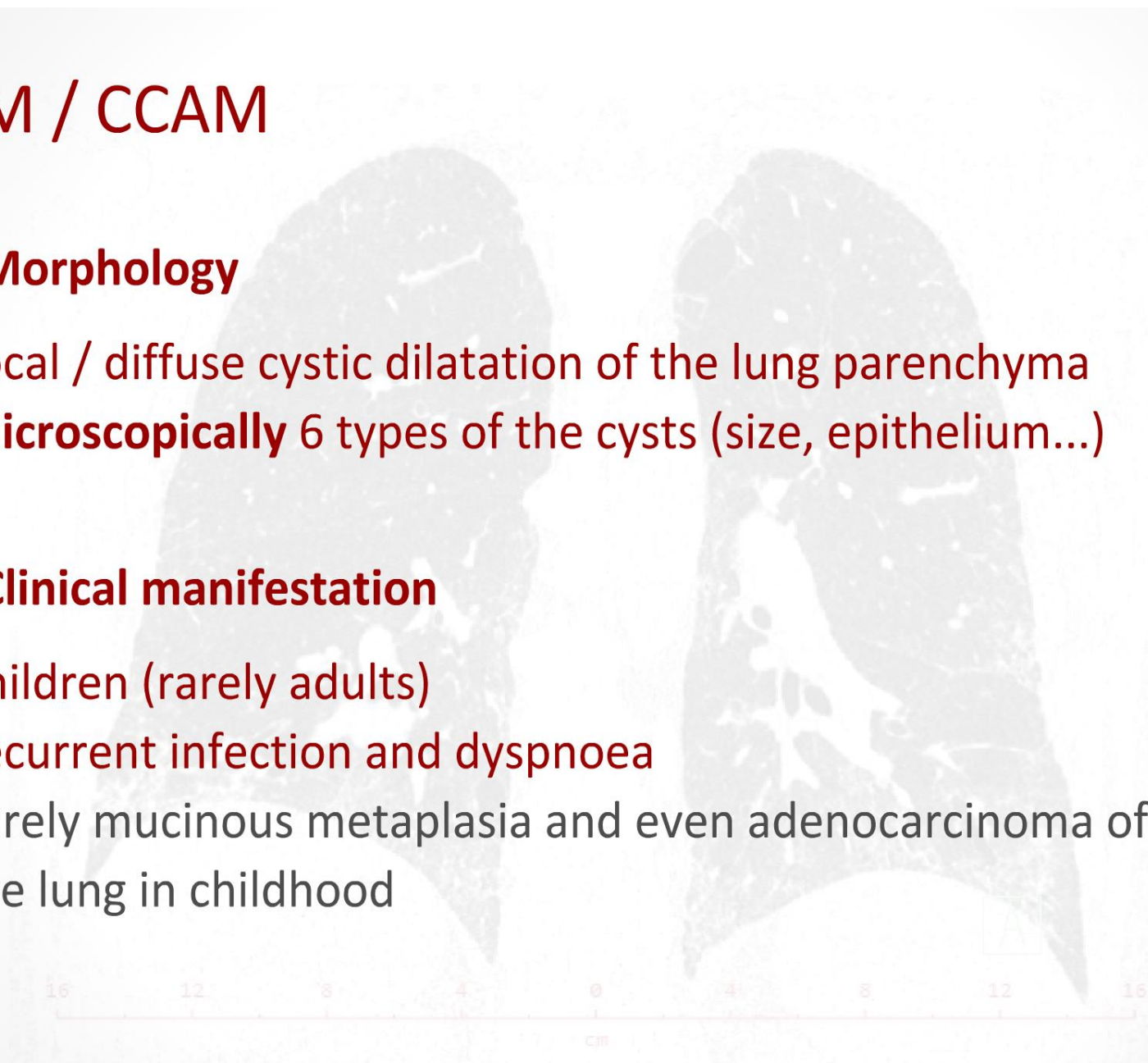
# CPAM / CCAM

## Morphology

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

## Clinical manifestation

- children (rarely adults)
- 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 development

## Developemant (pathogenesis)

- not communicating with **bronchial** tree
  - bronchiectasis / mucostasis are developed in time, unable to cough up the mucus
- blood supply from the aberrant **artery** from aorta
  - volume overload of the small circulation (pulmonary hypertension)

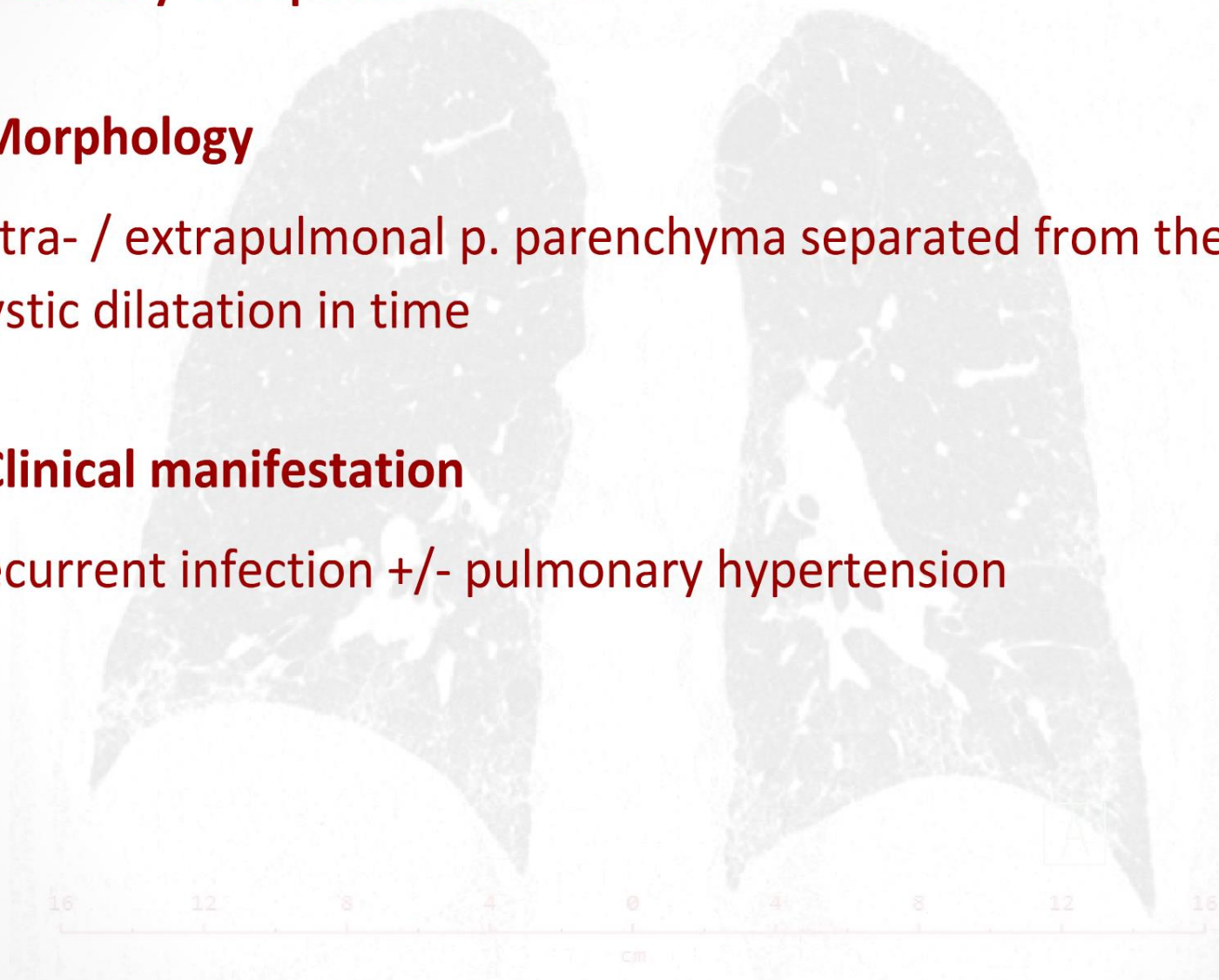
# Pulmonary sequestration

## Morphology

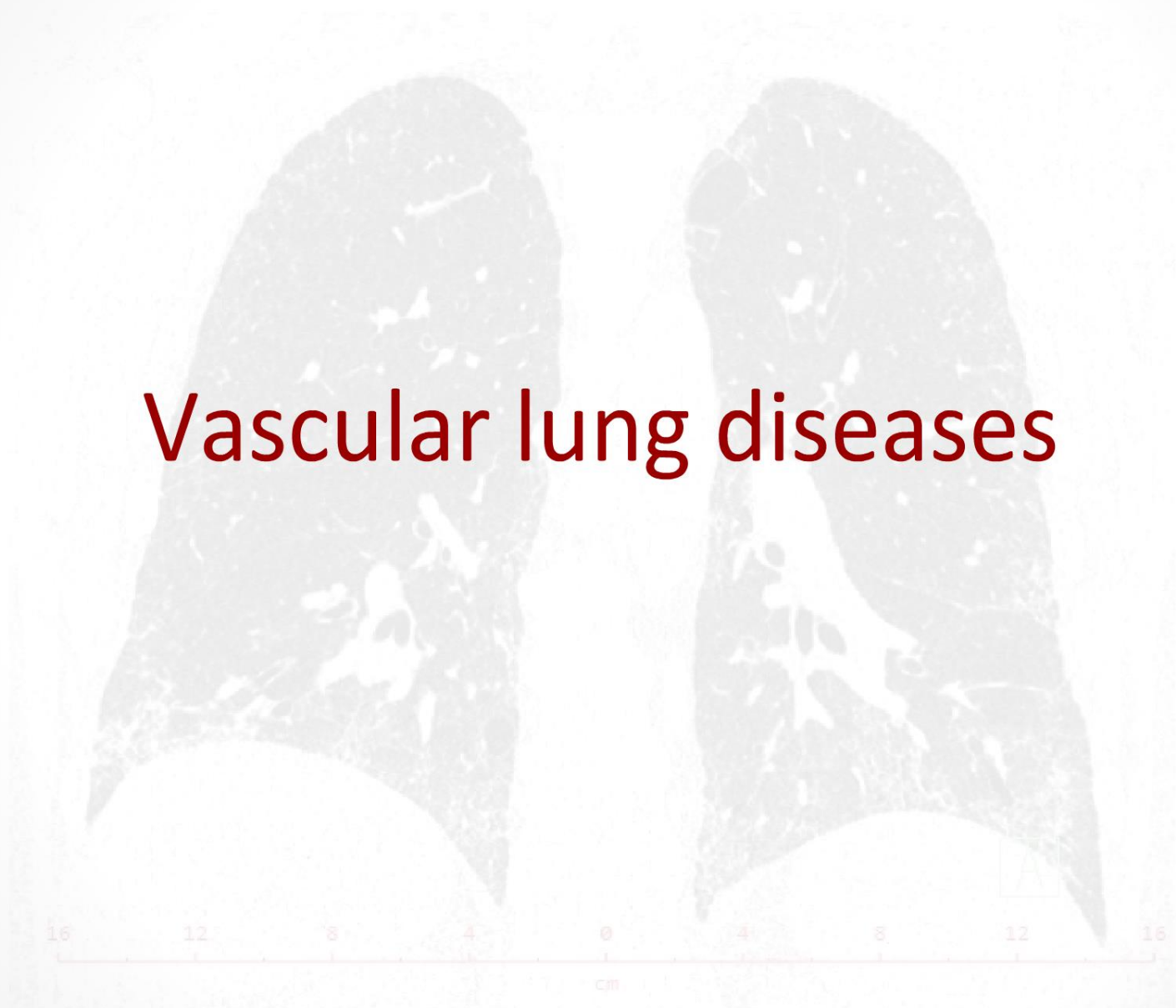
- intra- / extrapulmonal p. parenchyma separated from the lung
- cystic dilatation in time

## Clinical manifestation

- recurrent infection +/- 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)

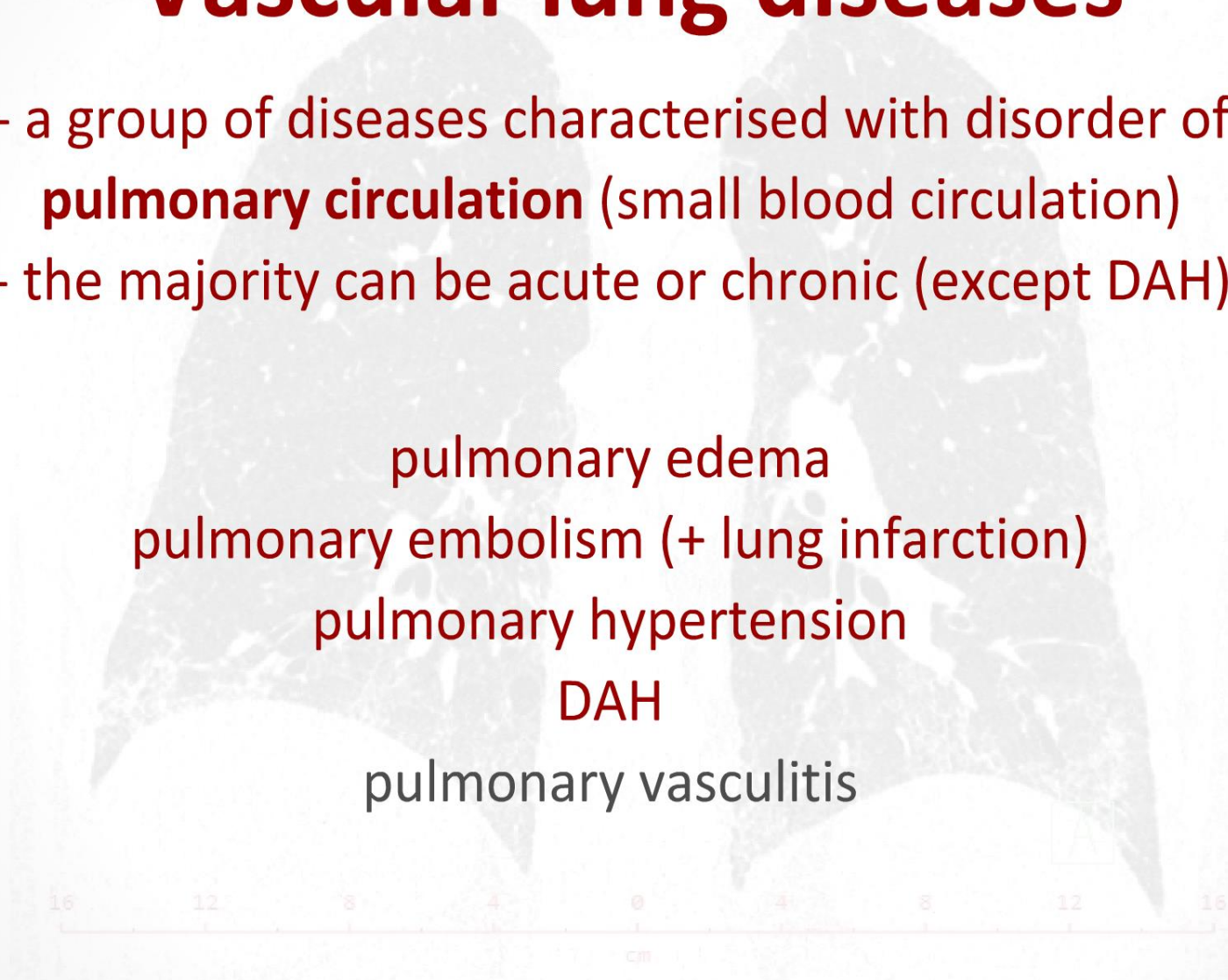
pulmonary edema

pulmonary embolism (+ lung infarction)

pulmonary hypertension

DAH

pulmonary vasculitis



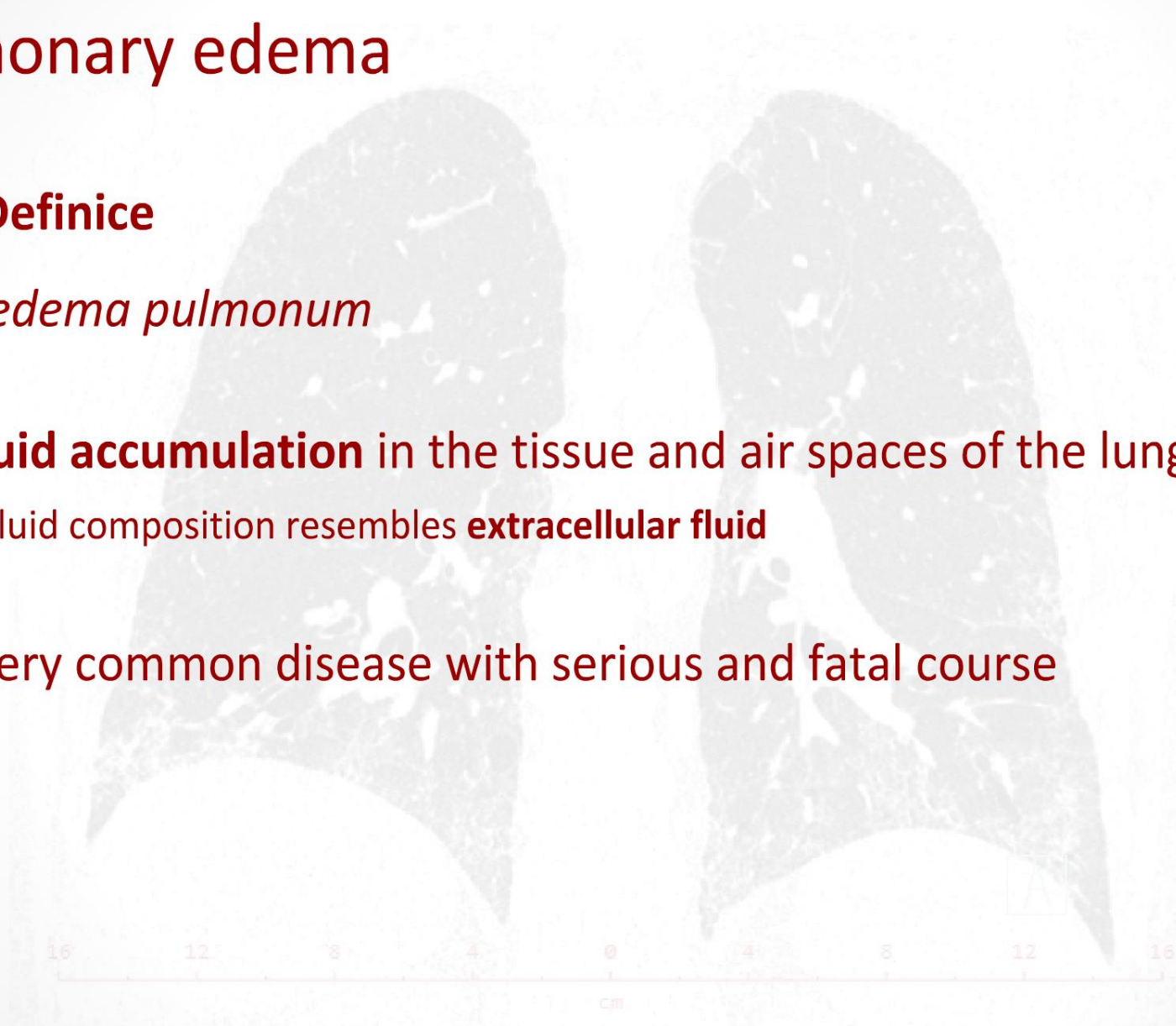


# Pulmonary edema



## 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**
  - hypertrophy of tunica media in **vessels**
  - **interstitium** undergoes fibrosis, which intensifies pulmonary hypertension
  - hypertension translates to the right **heart** (*cor translatum*)
- **complications** can develop
  - **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

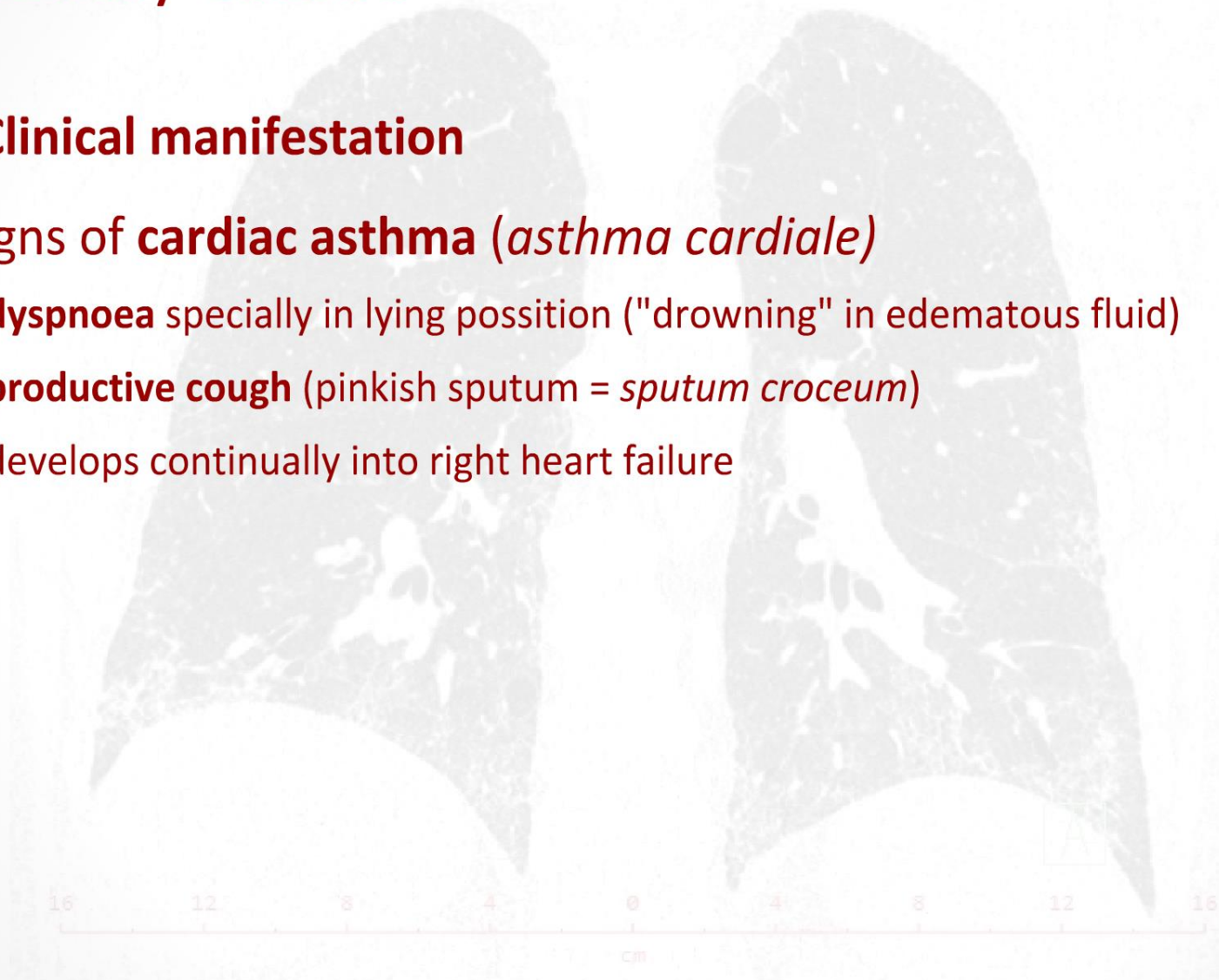




# Pulmonary edema

## ⊕ Clinical manifestation

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

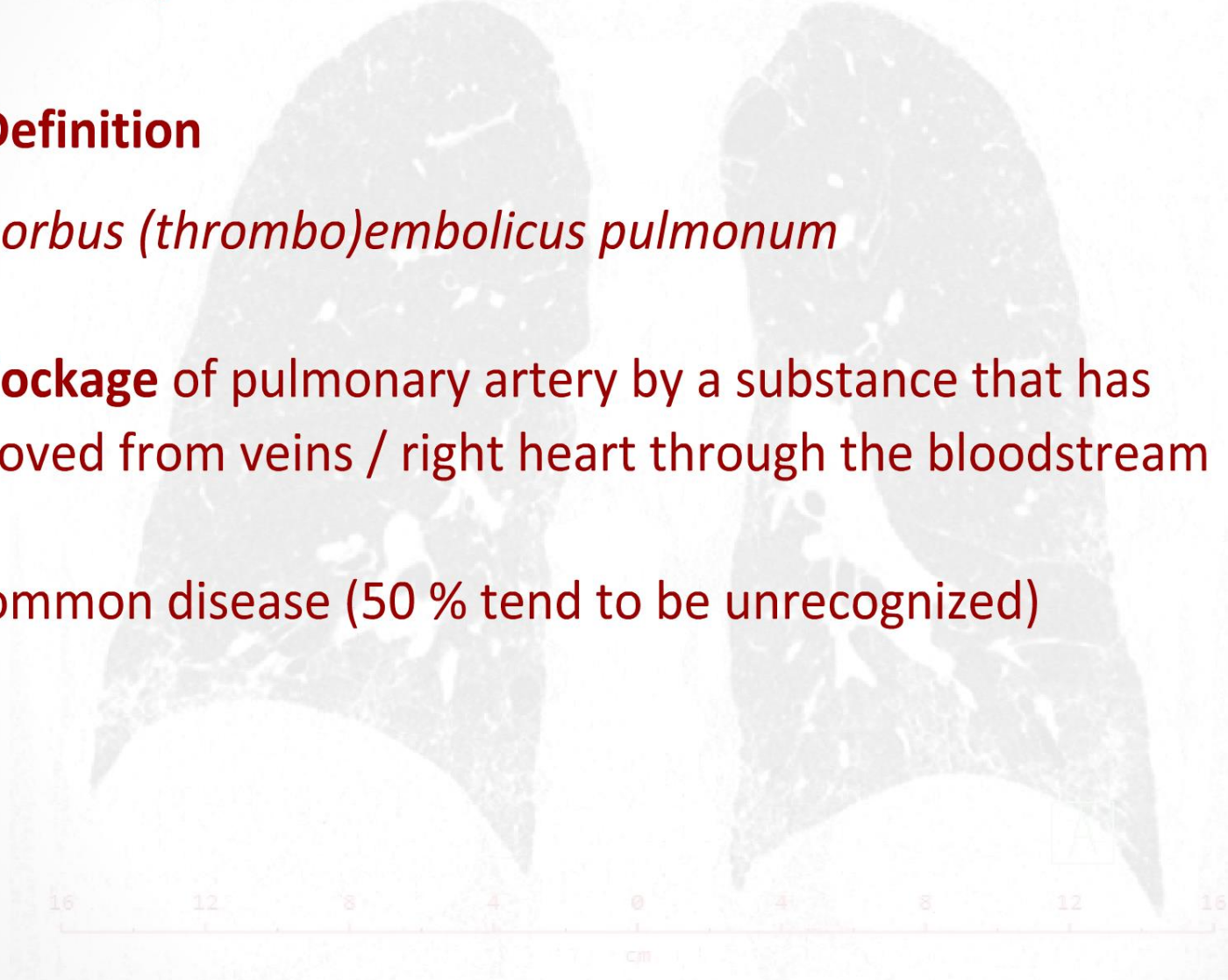


# Pulmonary embolism



## 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)

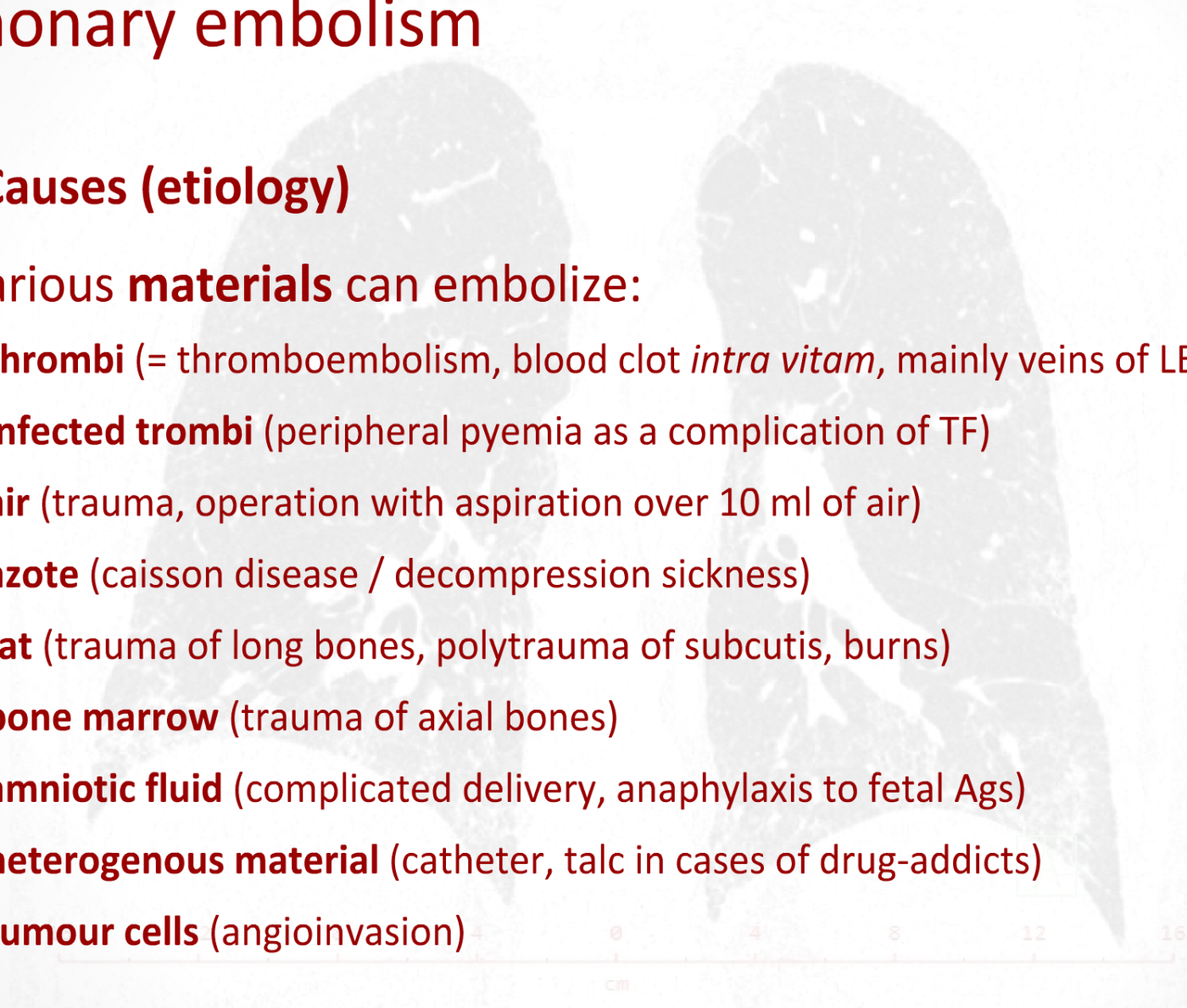


# Pulmonary embolism

## 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)

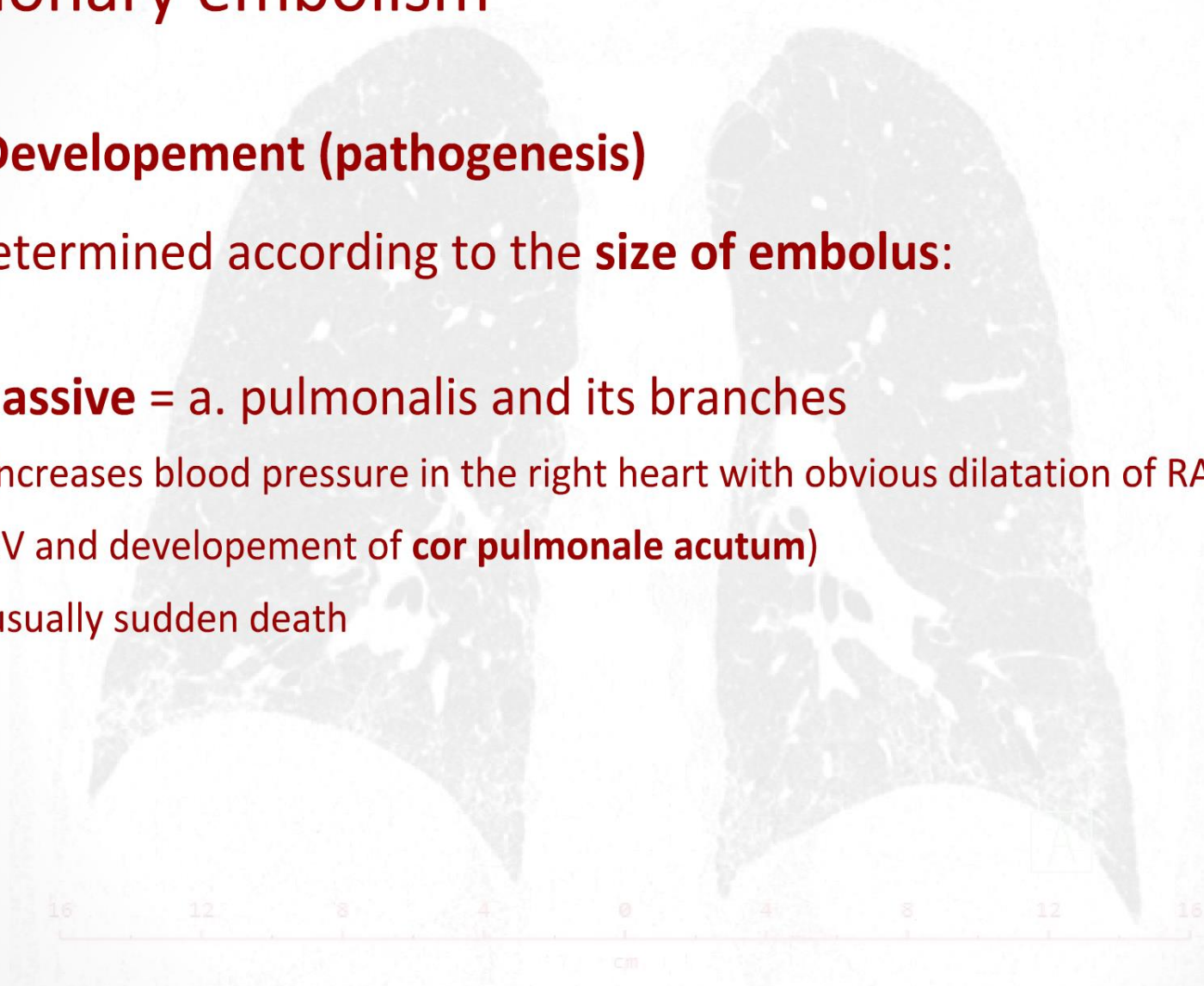


# Pulmonary embolism



## Development (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 development 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)
  - spenic **hemorrhagic necrosis** and fibrinous pleuritis (followed by scarring)
  - **complication** secondary anaerobic infection (pulmonary gangrene)

# Pulmonary embolism



## Developement (pathogenesis)

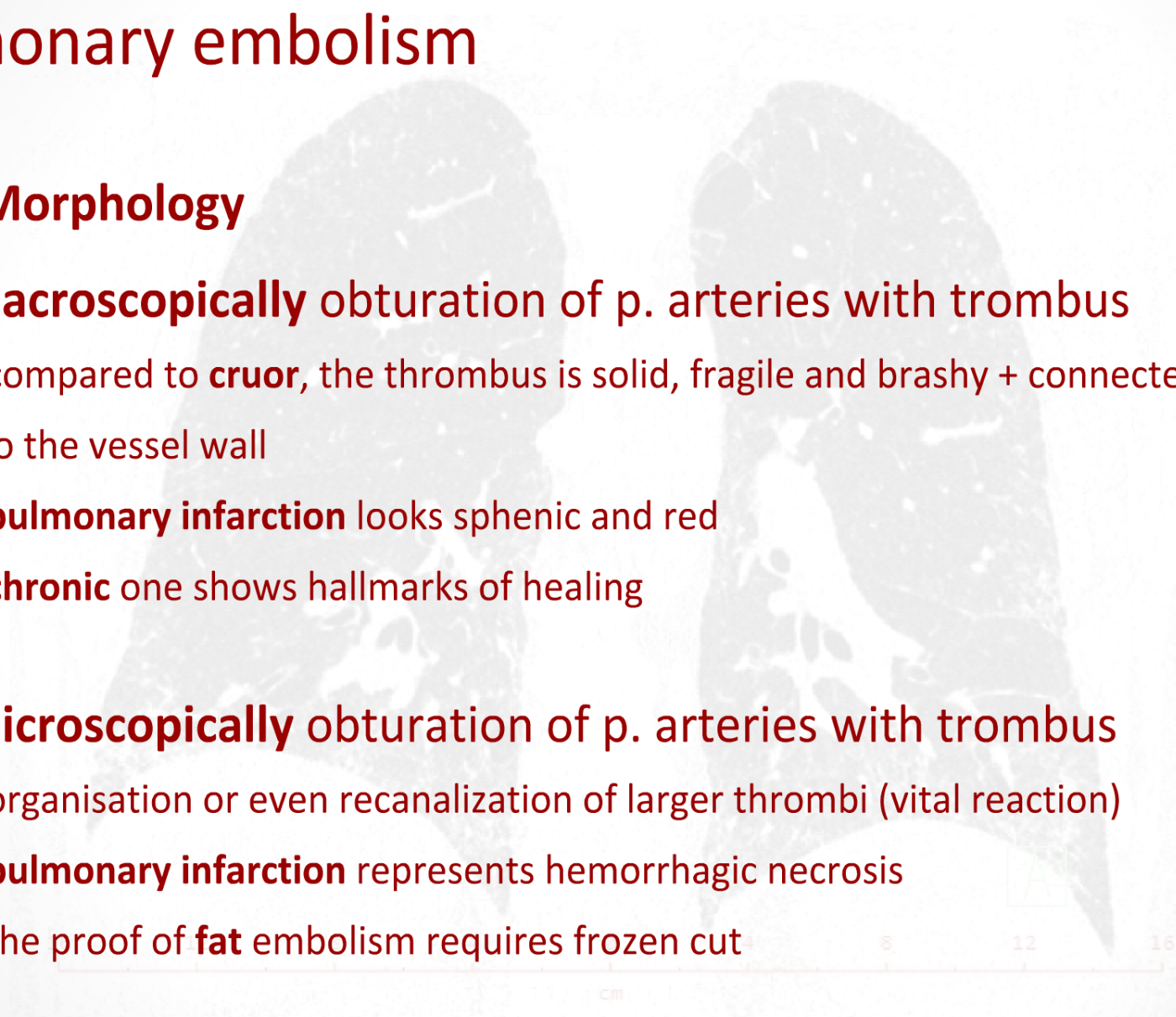
- determined according to the **size of embolus**:
- **successive** = capillaries (chronic, repeatedly)
  - gradual pulmonary hypertension without dilatation of RA + RV, but with hypertrophy = **cor pulmonale chronicum**)
  - heart failure (slow) and pulmonary infarction (numerous) are also possible

# Pulmonary embolism



## Morphology

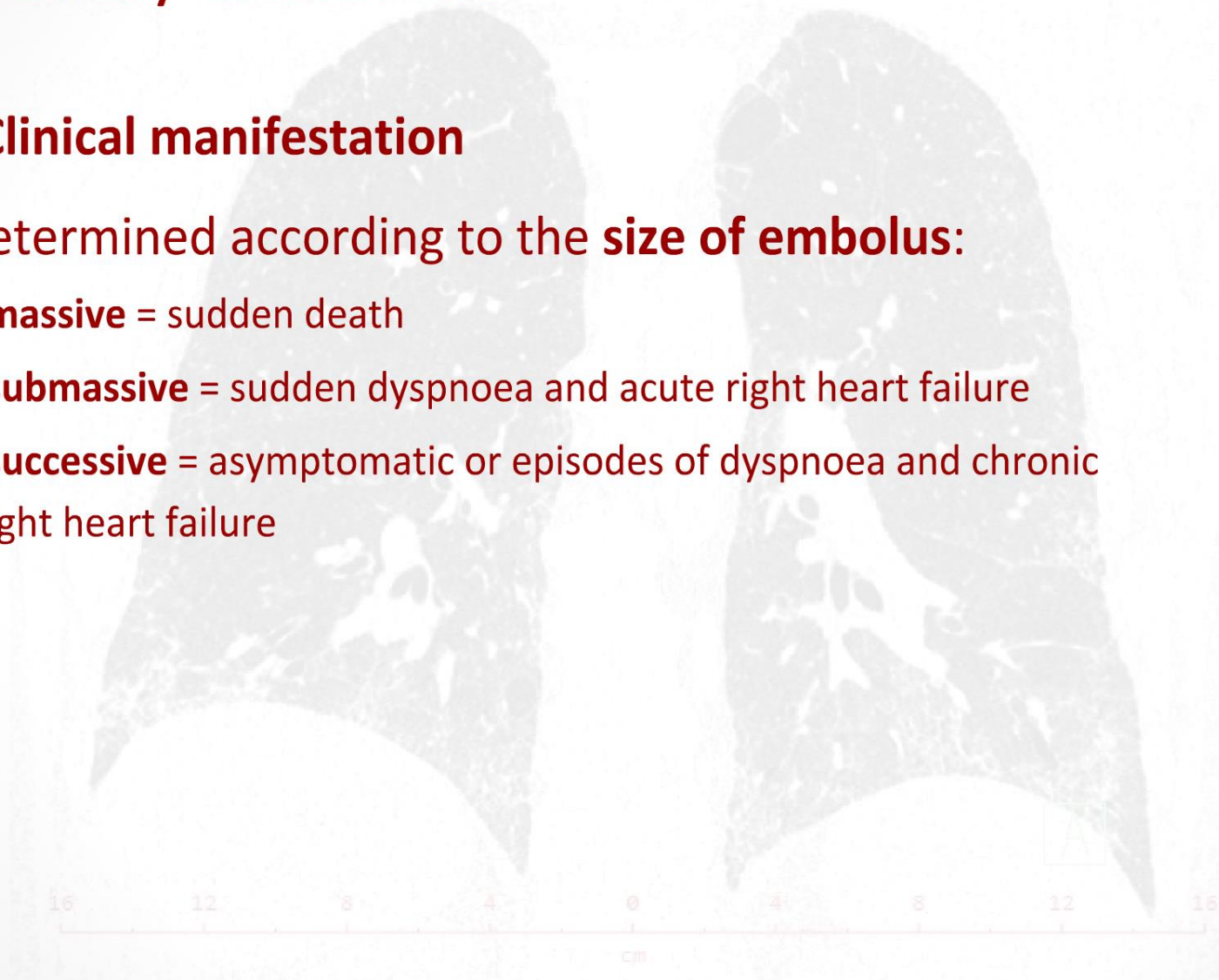
- **macroscopically** obturation of p. arteries with trombus
  - compared to **cruor**, the thrombus is solid, fragile and brashy + connected to the vessel wall
  - **pulmonary infarction** looks splenic and red
  - **chronic** one shows hallmarks of healing
- **microscopically** obturation of p. arteries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



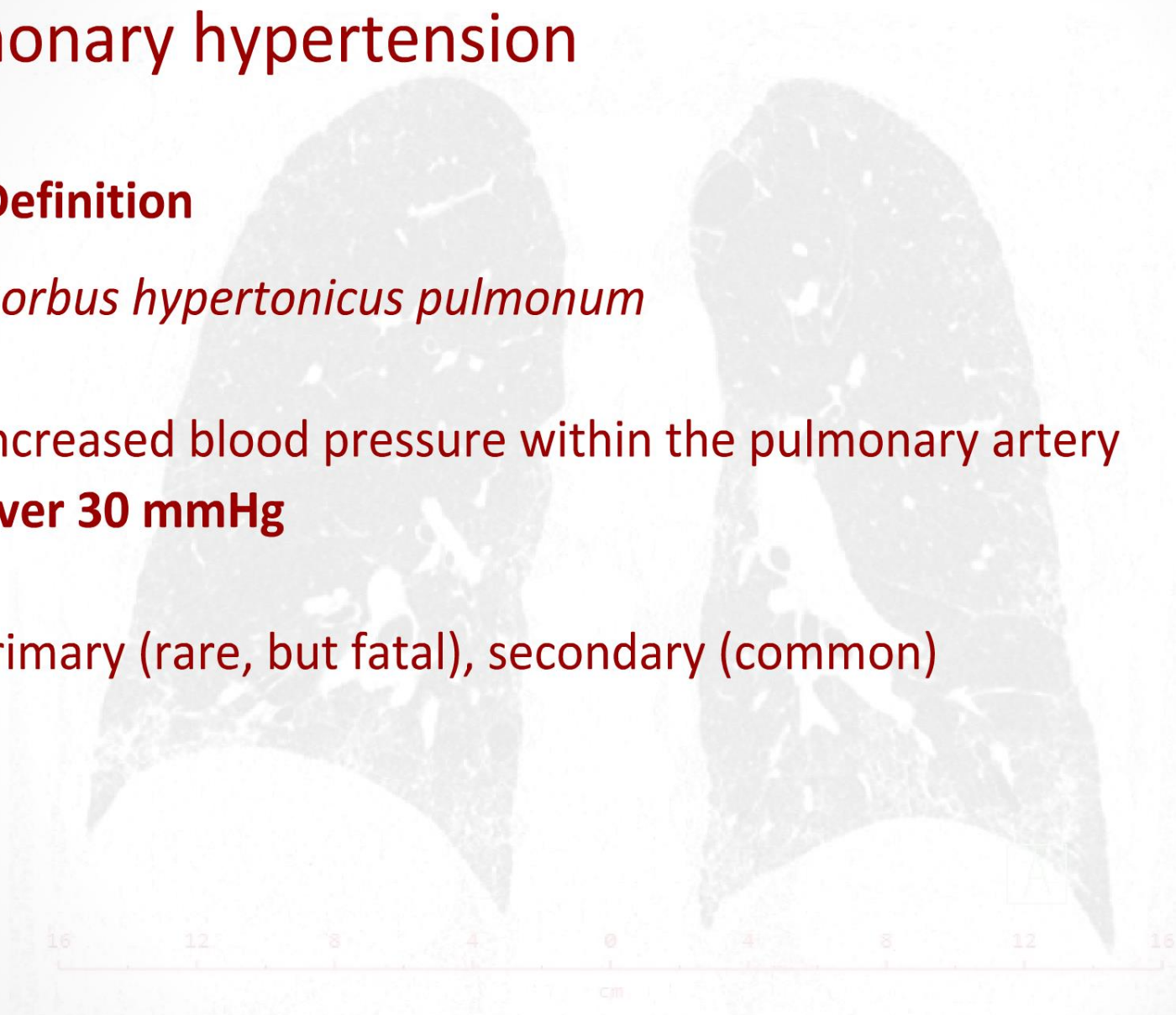


# Pulmonary hypertension



## Definition

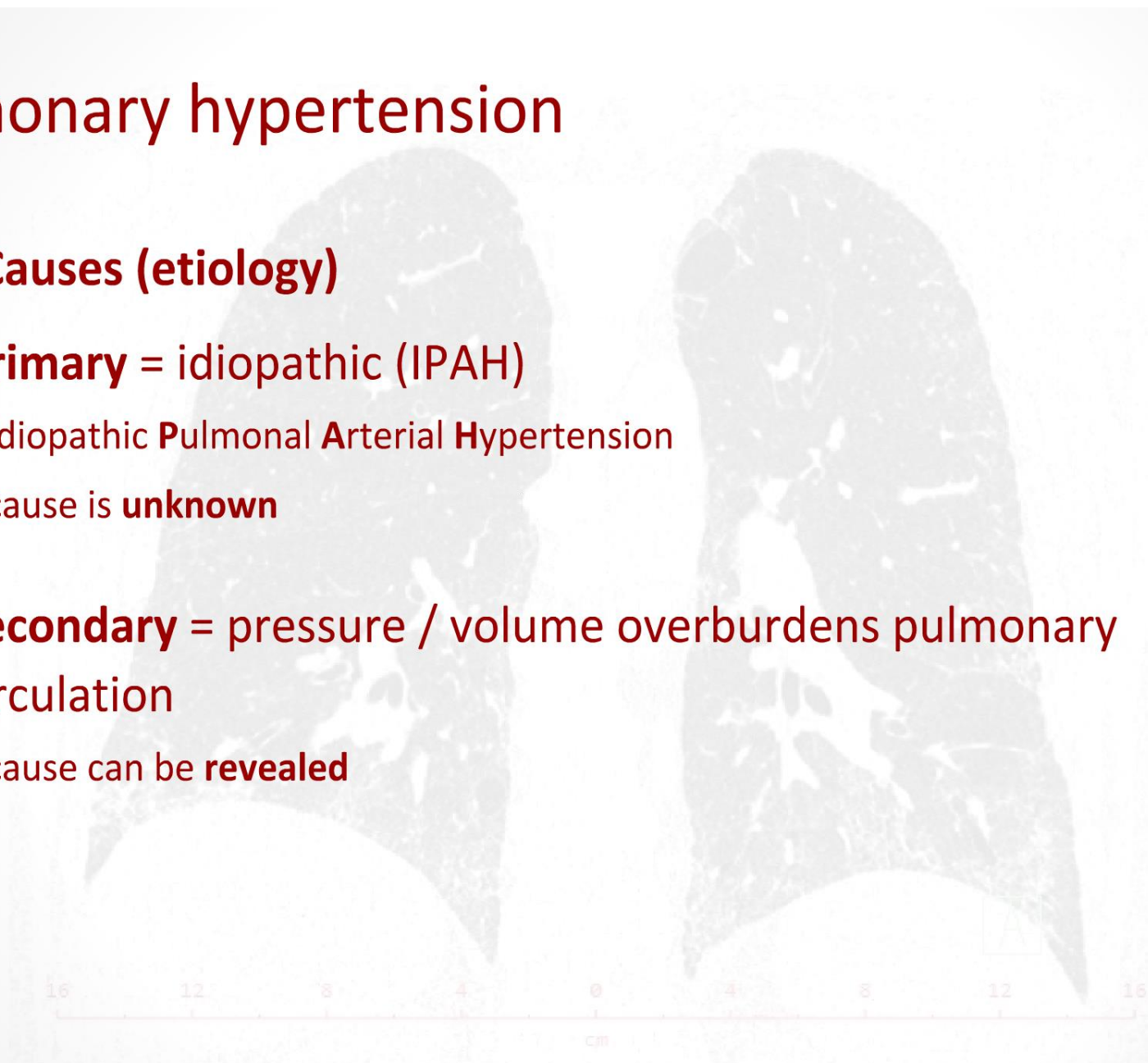
- *morbus hypertonicus pulmonum*
- increased blood pressure within the pulmonary artery  
**over 30 mmHg**
- primary (rare, but fatal), secondary (common)



# Pulmonary hypertension

## Causes (etiology)

- **primary** = idiopathic (IPAH)
  - Idiopathic Pulmonal Arterial Hypertension
  - 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 translutum, pulmonary veno-occlusive disease
- always leads to the overloading of RV and **cor pulmonale chronicum**

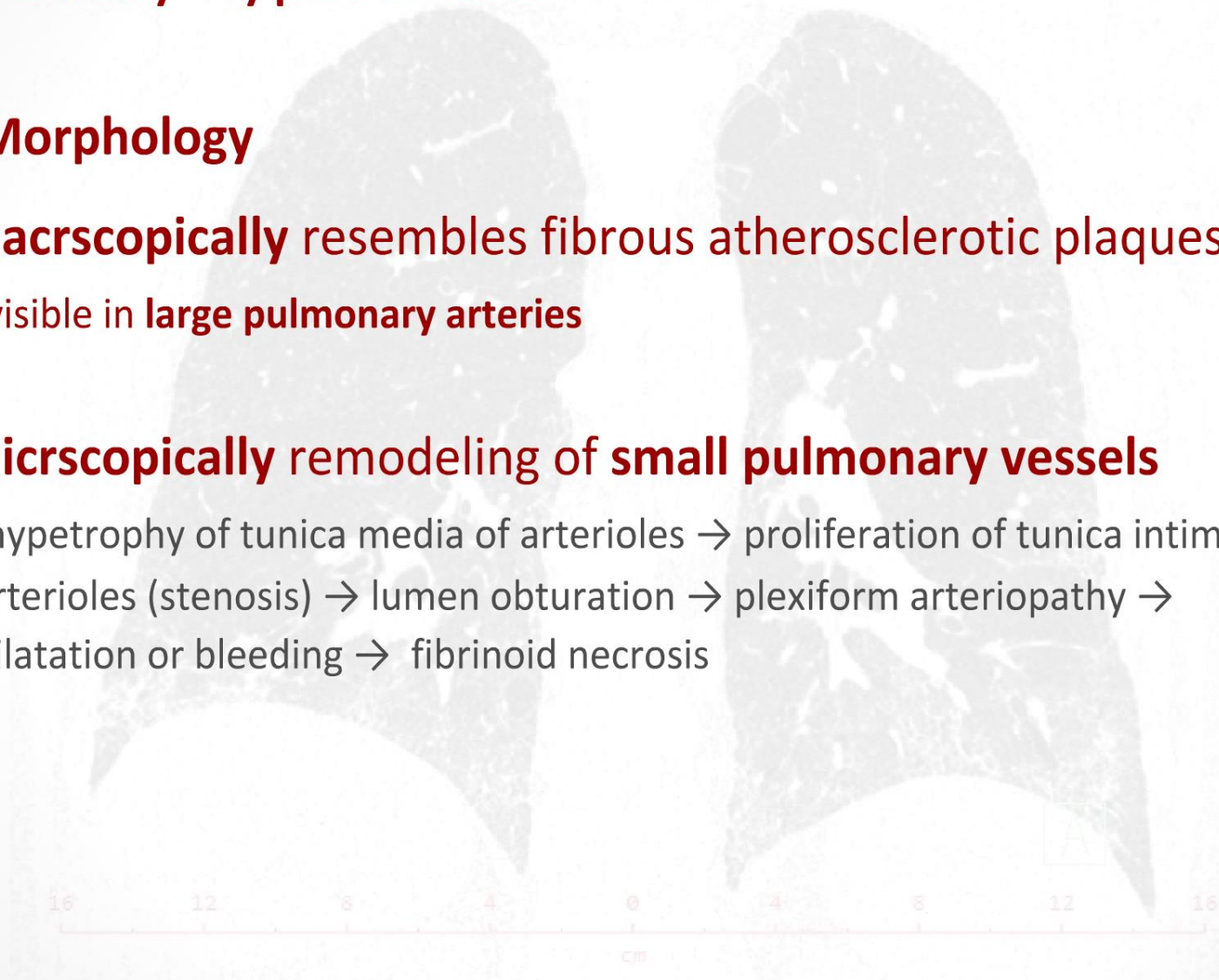


# Pulmonary hypertension



## Morphology

- **macroscopically** resembles fibrous atherosclerotic plaques
  - visible in **large pulmonary arteries**
- **microscopically** remodeling of **small pulmonary vessels**
  - hypertrophy of tunica media of arterioles → proliferation of tunica intima of arterioles (stenosis) → lumen obturation → plexiform arteriopathy → dilatation or bleeding → fibrinoid necrosis

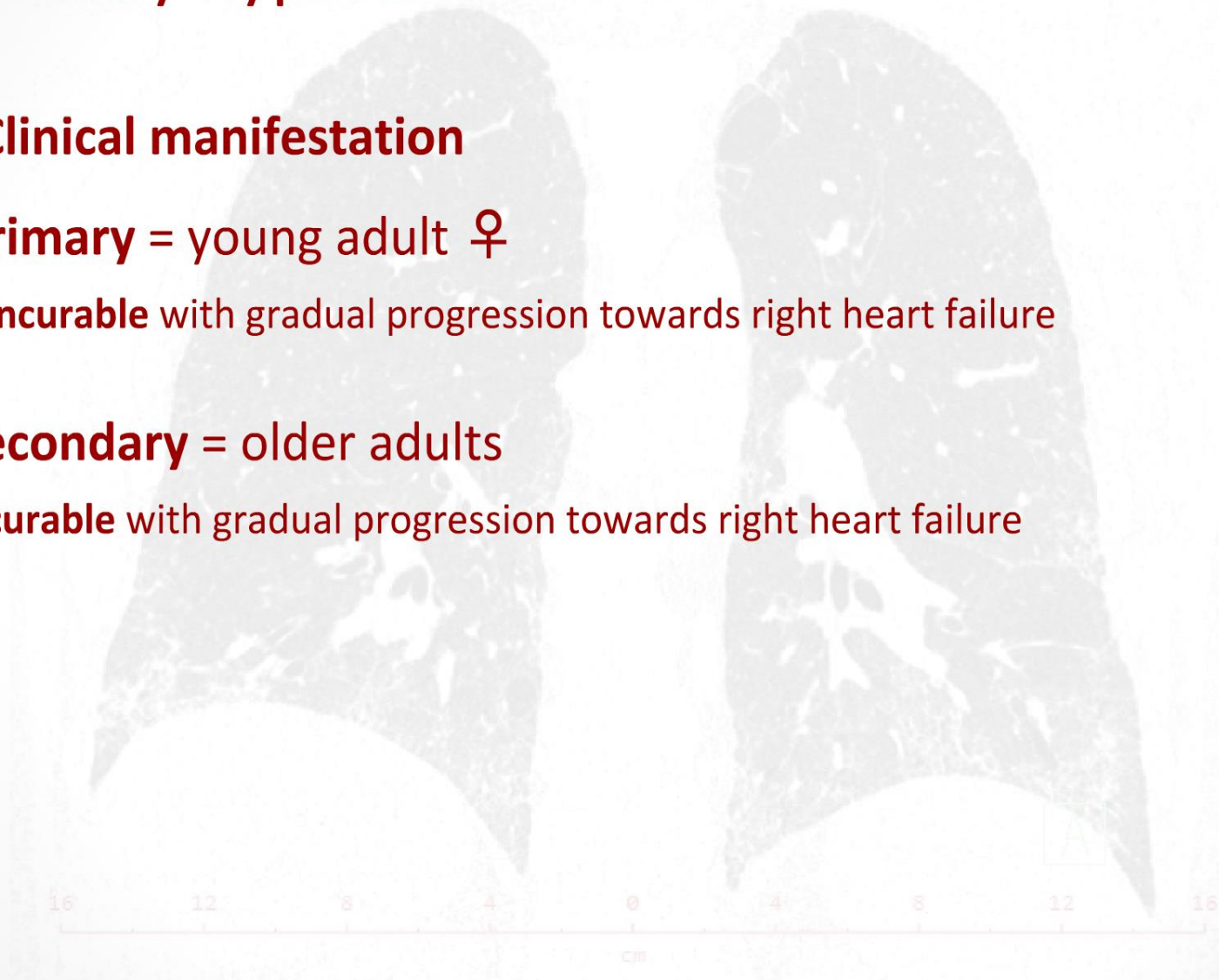




# Pulmonary hypertension

## ⊕ Clinical manifestation

- **primary** = young adult ♀
  - **incurable** with gradual progression towards right heart failure
- **secondary** = older adults
  - **curable** with gradual progression towards right heart failure

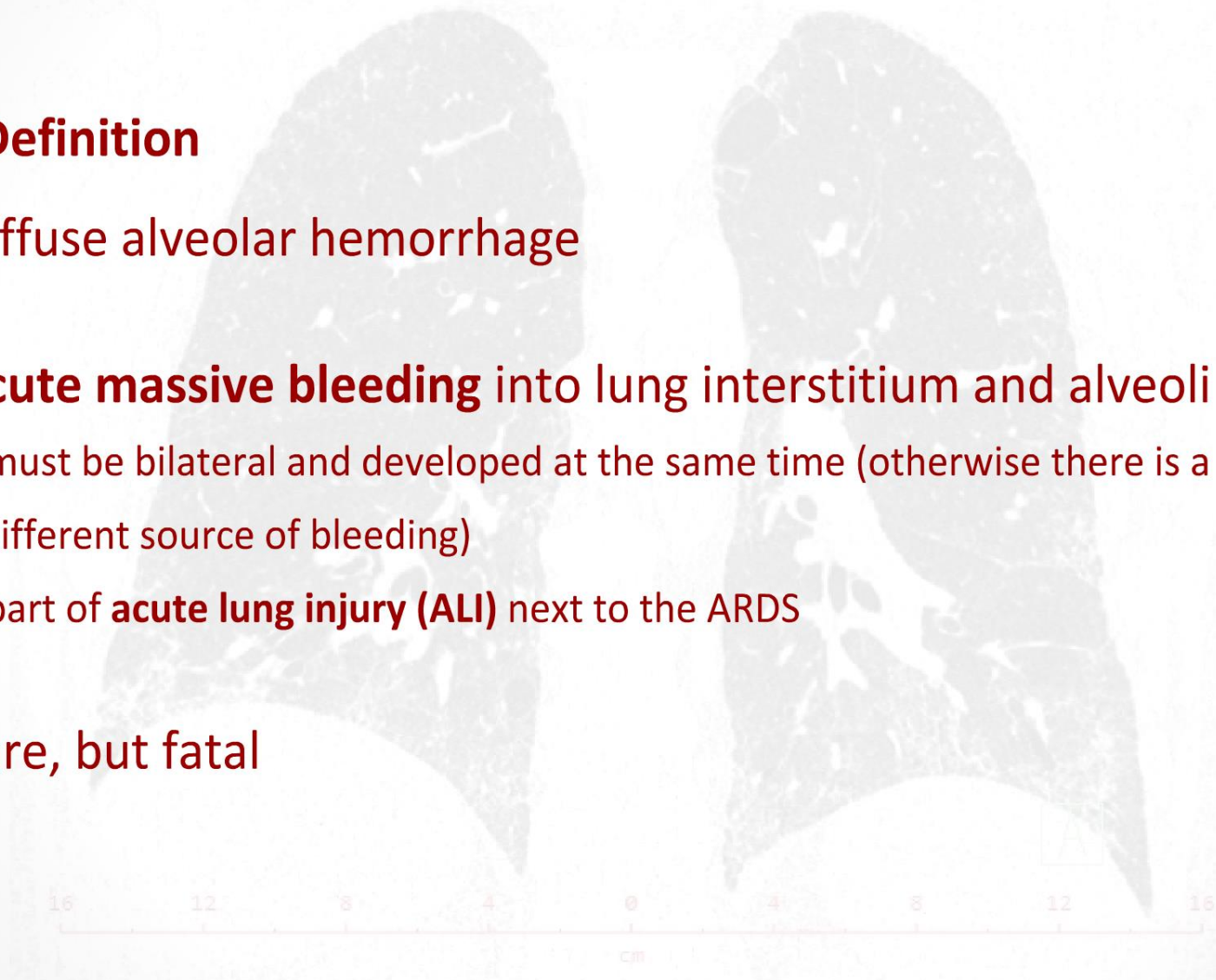


# DAH



## 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

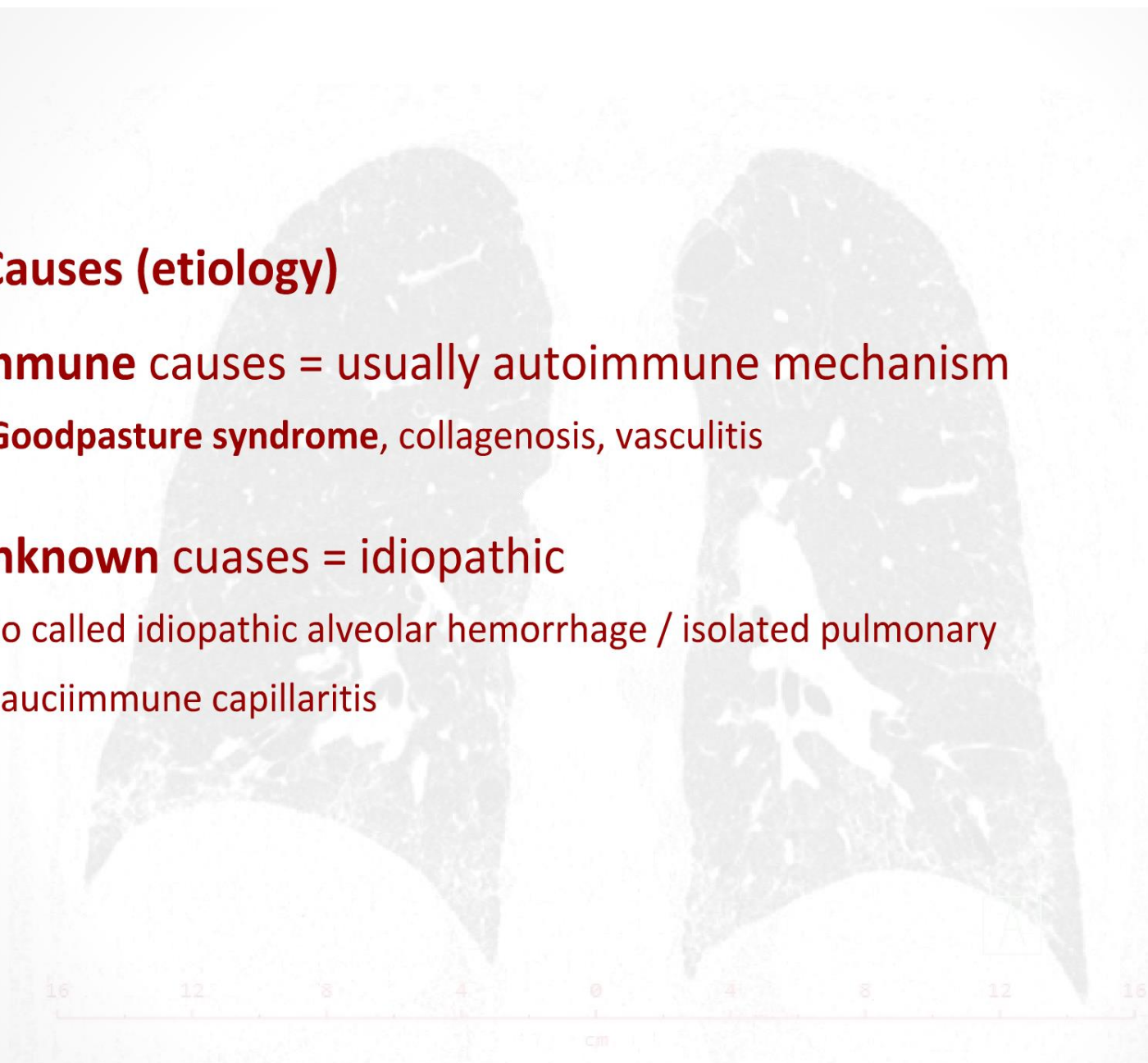


# DAH



## Causes (etiology)

- **immune** causes = usually autoimmune mechanism
  - **Goodpasture syndrome**, collagenosis, vasculitis
- **unknown** causes = idiopathic
  - so called idiopathic alveolar hemorrhage / isolated pulmonary pauciimmune capillaritis



# DAH



## Development (pathogenesis)

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

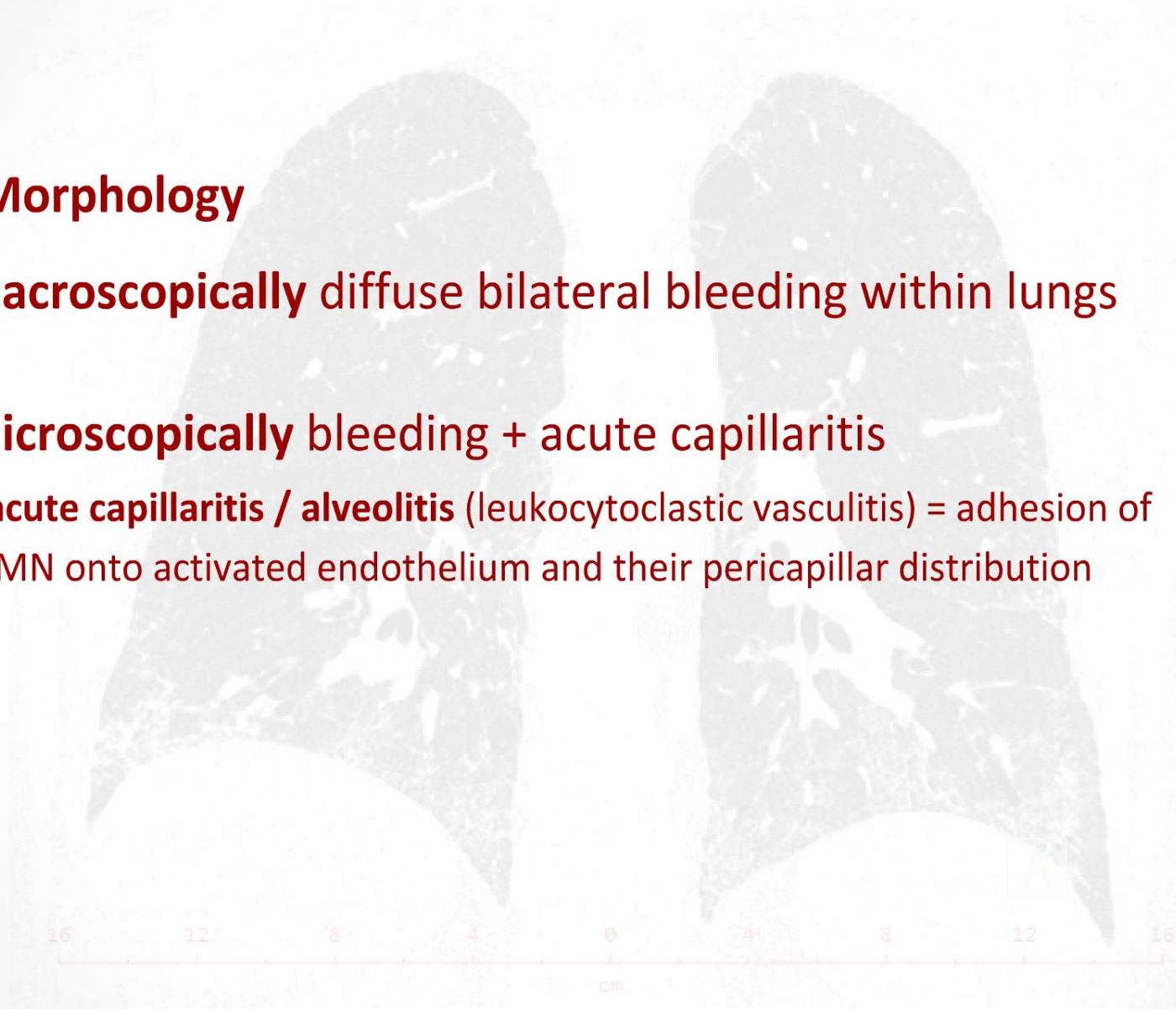




# DAH

## 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

- always **severe condition**

- dyspnoea with **hemoptysis** and life-threatening RI



**Atelectasis / collapsed lung**

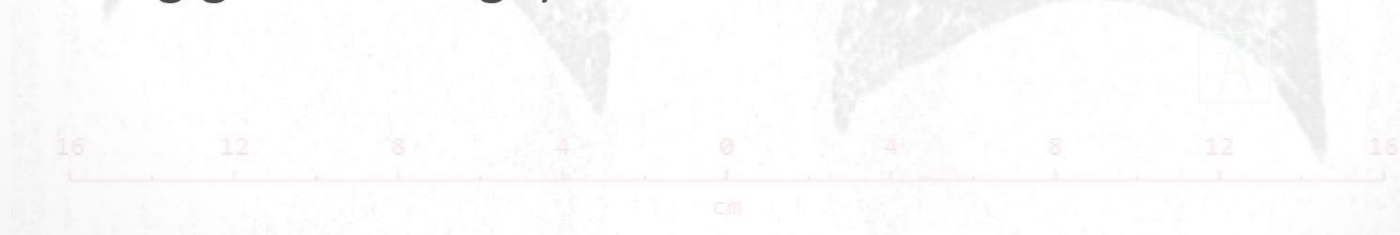


# Atelectasis / collapsed lung



## 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 ascending bronchus 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)

- alveolar collapse after 1<sup>st</sup> inbreath of immature newborns

# Atelectasis / collapsed lung

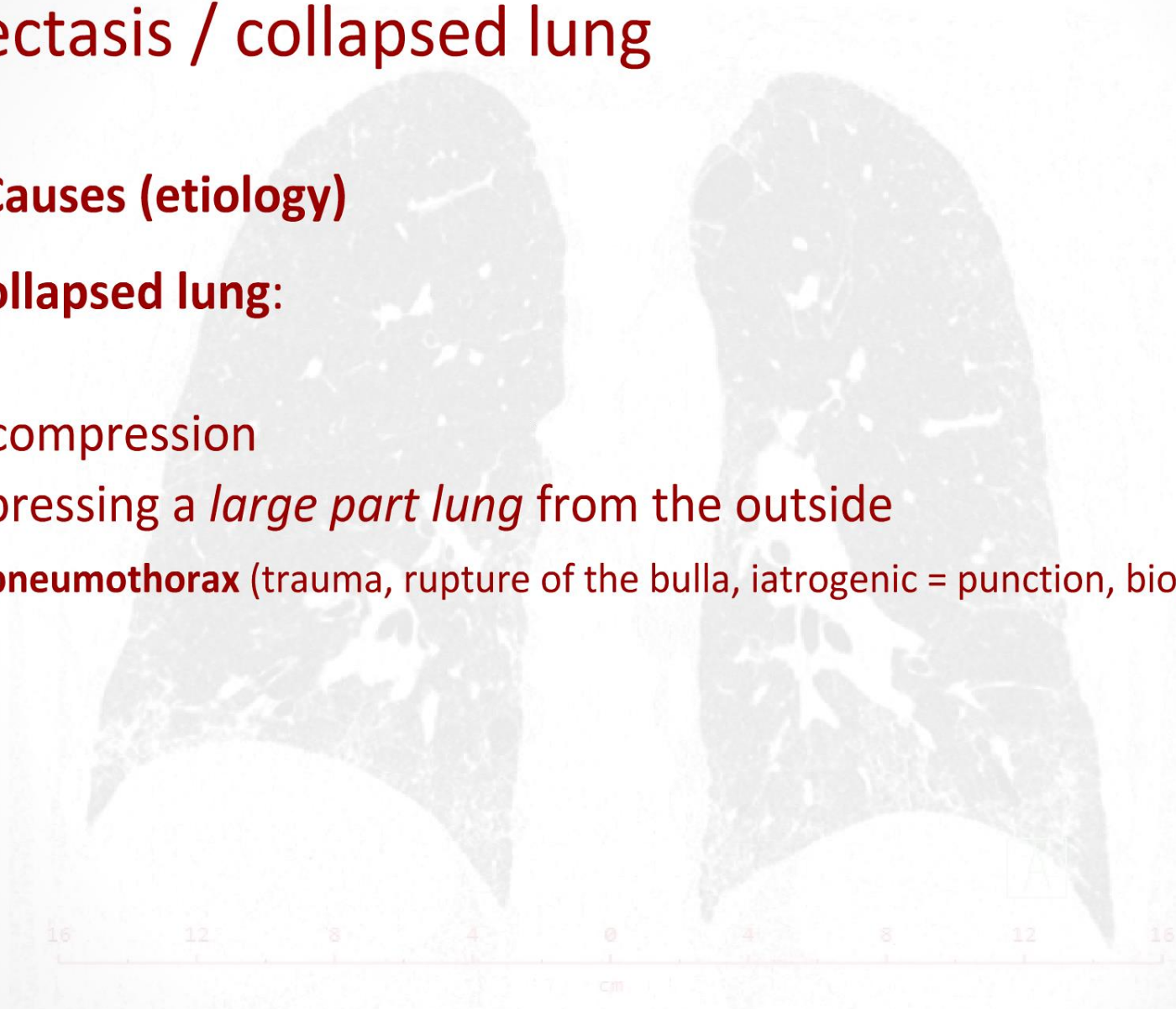
## 🔑 Causes (etiology)

- collapsed lung:

### 1) compression

- pressing a *large part lung* from the outside

- **pneumothorax** (trauma, rupture of the bulla, iatrogenic = puncture, biopsy)



# Atelectasis / collapsed lung



## Developement (pathogenesis)

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



# Atelectasis / collapsed lung

## Morphology

- **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, mediastinal shift
  - water test in newborns (except from *pulmo spumousus*)
- **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 application into pleural cavity
- **acute**
  - inflation of the parenchyma after elimination of the cause (reversible)
- **chronic**
  - fibrotisation (induration) of the lung parenchyma (irreversible)

Thank you for attention.



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