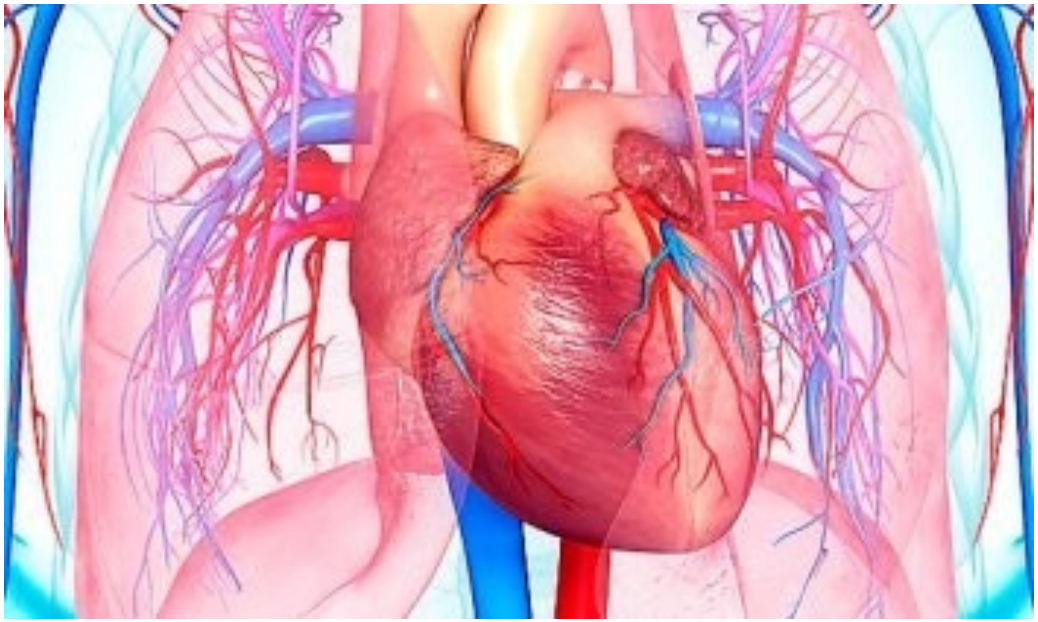
PULMONARY DISEASES OF VASCULAR ORIGIN



MARAM ABDALJALEEL, MD.
SCHOOL OF MEDICINE
THE UNIVERSITY OF JORDAN

https://www.europeanlung.org/en/lung-disease-and-information/lung-diseases/pulmonary-vascular-disease

PULMONARY DISEASES OF VASCULAR ORIGIN

- Pulmonary Embolism, Hemorrhage, and Infarction
- Pulmonary Hypertension
- Diffuse Alveolar Hemorrhage Syndromes

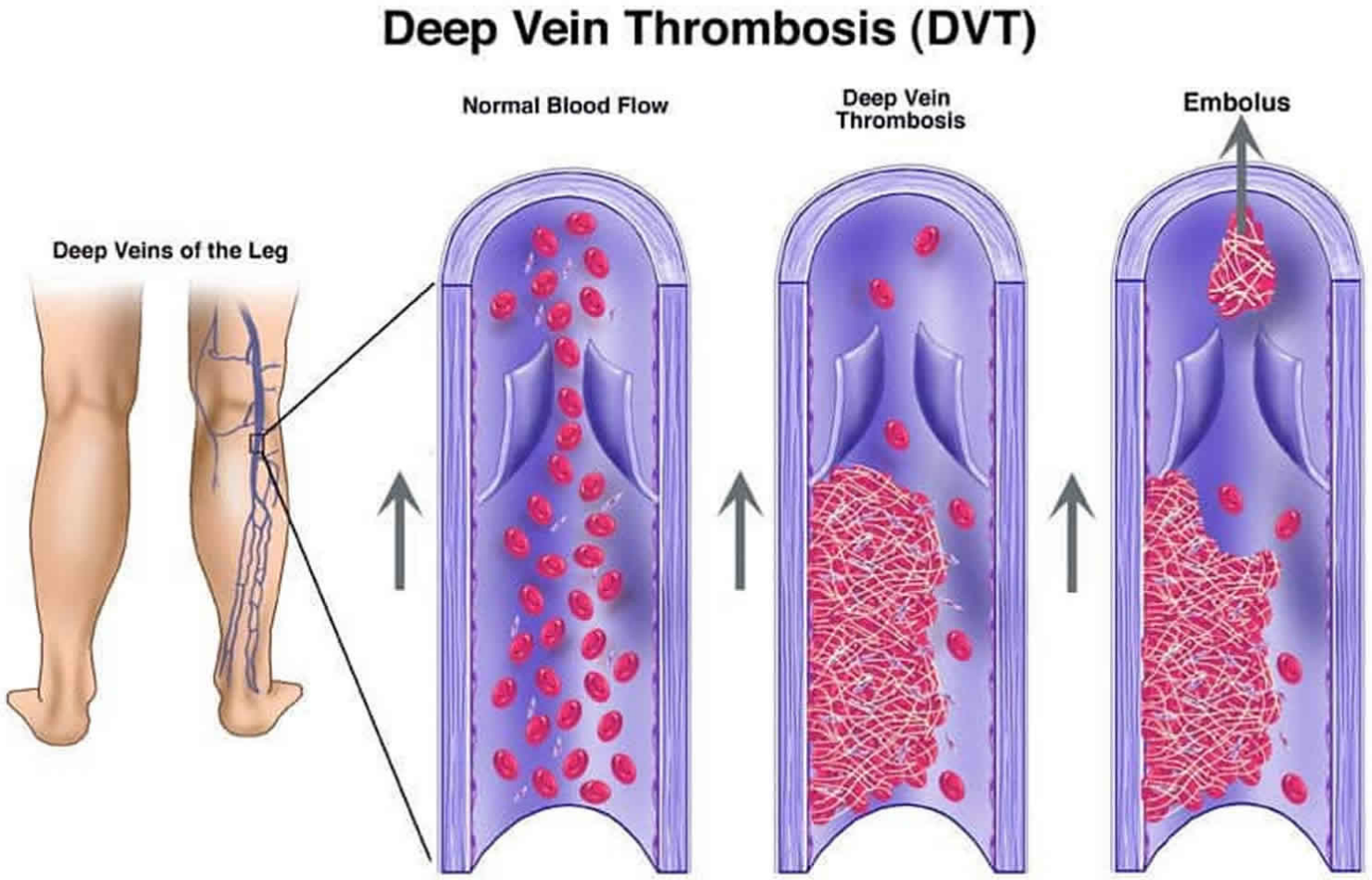
PULMONARY EMBOLISM:

- **Thromboembolism**
- **Nonthrombotic pulmonary emboli**

THROMBOEMBOLISM

- Almost all large pulmonary artery thrombi are embolic in Origin.

- >95% of PE arise from thrombi within the large deep veins of the legs, most often popliteal vein and larger veins above it.



RISK FACTORS FOR VENOUS THROMBOSIS:

1. prolonged bed rest (immobilization of the legs)
2. Surgery (orthopedic surgery on the knee or hip)
3. severe trauma (burns or multiple fractures)
4. congestive heart failure
5. in women, the period around parturition or the use of OCPs (high estrogen content)
6. disseminated cancer
7. primary disorders of hypercoagulability (factor V Leiden)

CONSEQUENCES

1. increase in pulmonary artery pressure and vasospasm

2. ischemia of the downstream pulmonary parenchyma.

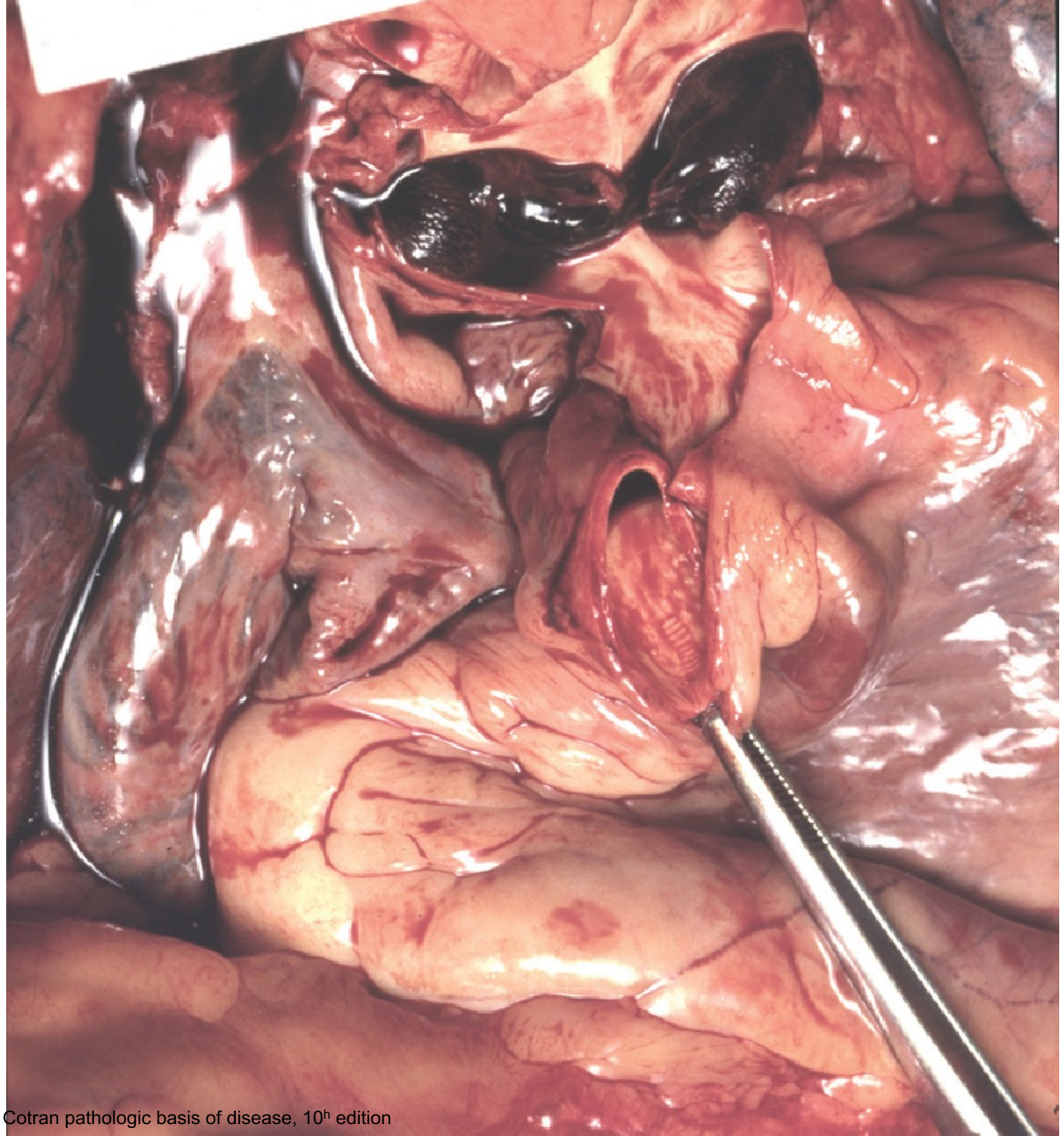
CONSEQUENCES:

- **depend mainly on:**

1- size of the embolus:

- large embolus may embed in the main pulmonary artery or its major branches or lodge at the bifurcation as a saddle embolus
- Smaller emboli become impacted in medium-sized and small-sized pulmonary arteries.

2- the cardiopulmonary status of the patient.



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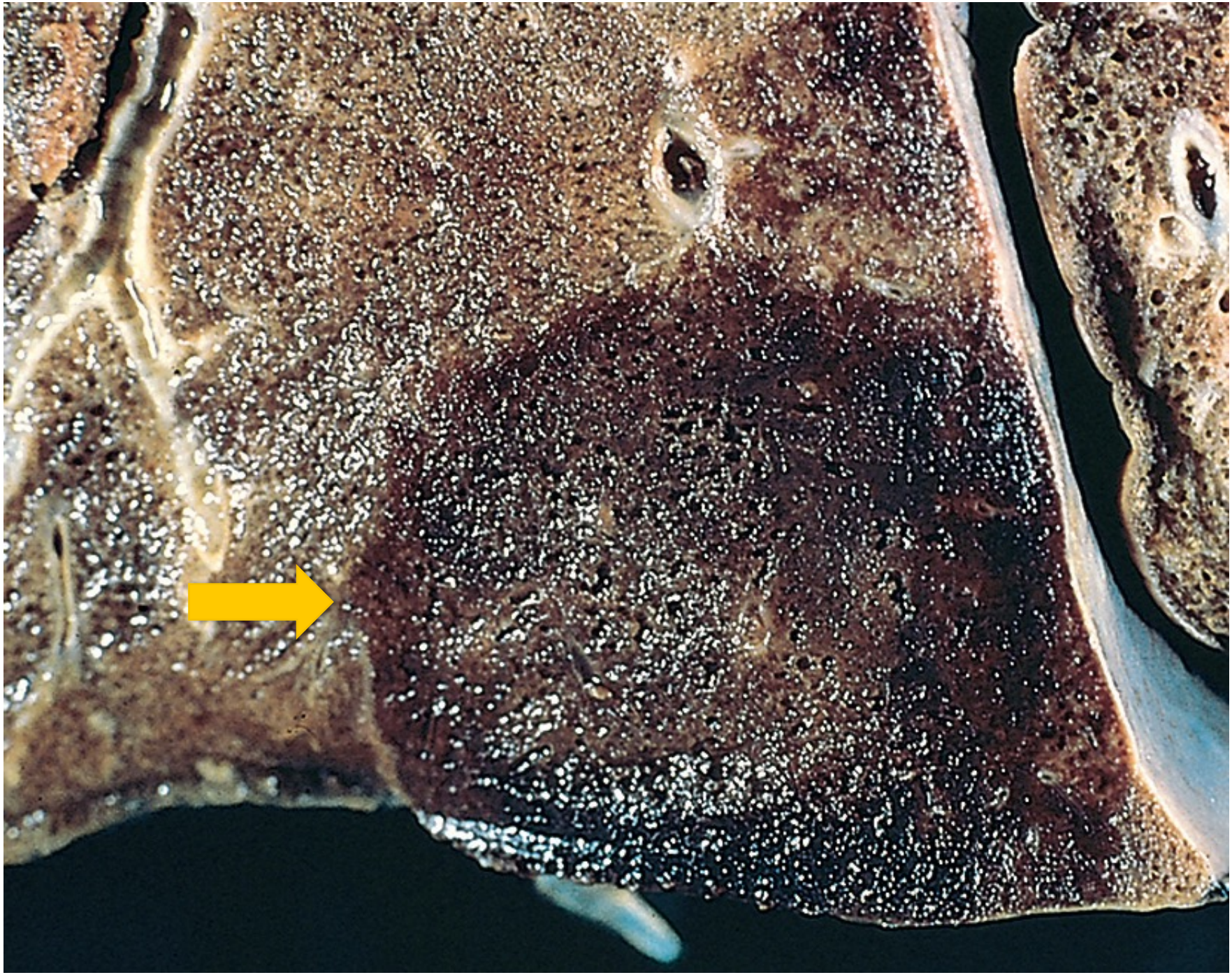
MORPHOLOGY:

- **No morphologic alternations**: large emboli
- **alveolar hemorrhage**: Smaller emboli
- **infarction :**
  - compromised cardiovascular status (congestive heart failure)
  - The more peripheral the embolic occlusion, the higher the risk for infarction.
    - ¾ lower lobes & >50% multiple.
  - wedge-shaped, with their base at the pleural surface and the apex pointing toward the hilus of the lung.

PULMONARY INFARCTS

- Typically, hemorrhagic with red-blue areas of coagulative necrosis in the early stages
- The adjacent pleura surface is covered by fibrinous exudate
- The occluded vessel is located near the apex of the infarcted area.
- The red cells begin to lyse within 48 hrs→ red-brown as hemosiderin is produced → fibrous replacement begins at the margins as a gray-white peripheral zone →scar.



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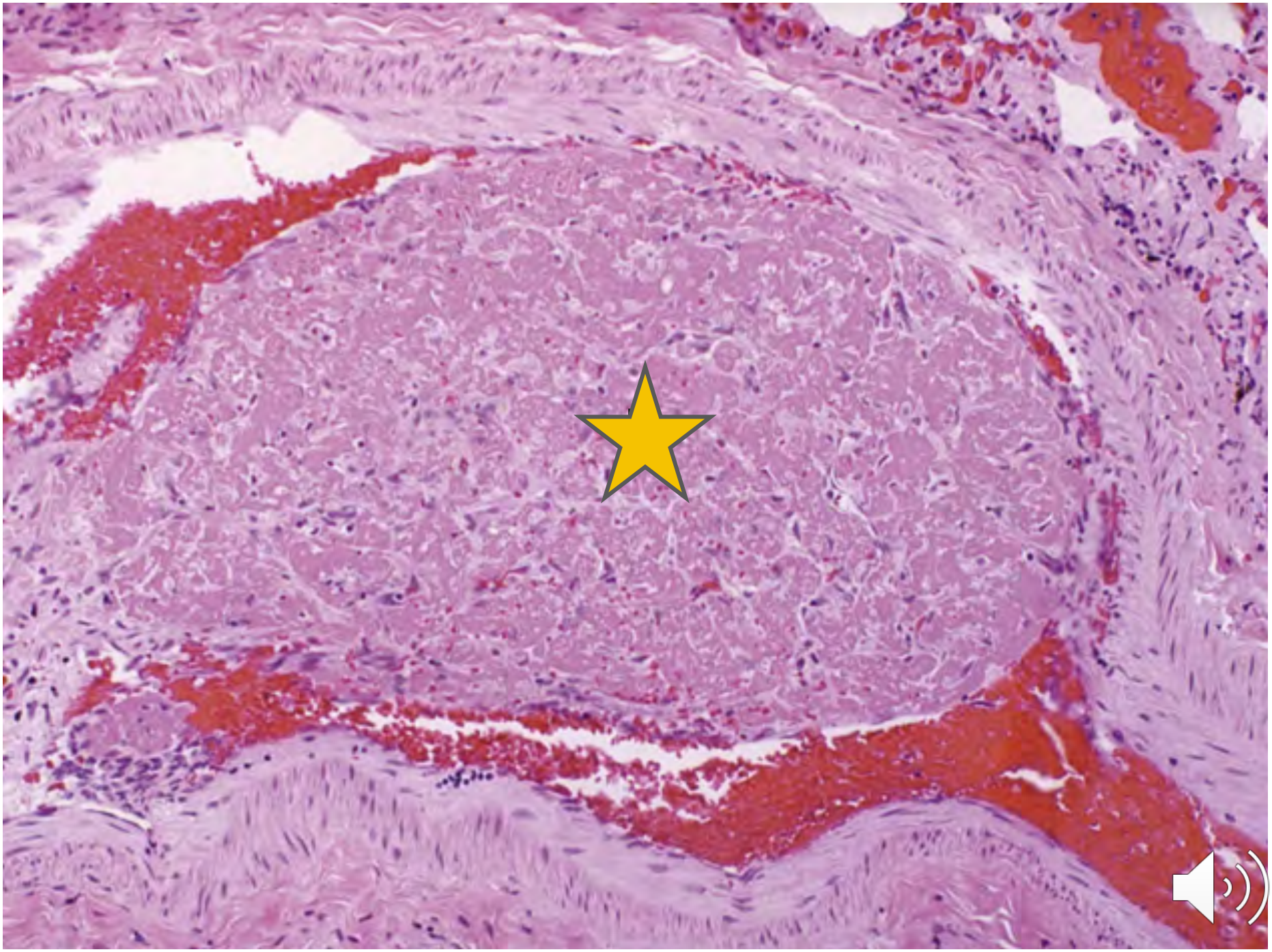


CLINICAL FEATURES

- 60% - 80% →clinically silent
- Small emboli
- embolic mass is rapidly removed by fibrinolytic activity.

- 5%→ death, acute right-sided heart failure, or cardiovascular collapse.
  - As in Massive pulmonary embolism: >60% of the total pulmonary vasculature is obstructed by a large embolus or multiple small emboli.

CLINICAL FEATURES

- 10-15%→ dyspnea
- Obstruction of small to medium pulmonary branches→ pulmonary infarction

- <3% →progressively worsening dyspnea
  - recurrent showers of emboli leading to pulmonary hypertension, chronic right-sided heart failure, and pulmonary vascular sclerosis.

MANAGEMENT:

- Prophylactic therapy: anticoagulation, early ambulation, elastic stockings, intermittent pneumatic calf compression, and isometric leg exercises for bedridden patients.

- anti-coagulation therapy for patients who develop pulmonary embolism

- thrombolytic therapy for hemodynamically unstable pts with massive pulmonary embolism

NONTHROMBOTIC PULMONARY EMBOLI:

- uncommon but potentially lethal

- such as:
  - air, fat, amniotic fluid embolism,
  - foreign body embolism in intravenous drug abusers
  - Bone marrow embolism:
    - the presence of hematopoietic and fat elements within a pulmonary artery
    - after massive trauma and in patients with bone infarction secondary to sickle cell anemia

PULMONARY HYPERTENSION

- defined as pressures of **25 mm Hg or more at rest**

- may be caused by a **decrease** in the cross-sectional area of the pulmonary vascular bed or by **increased** pulmonary vascular blood flow.

CLASSIFIED AS FOLLOWING:

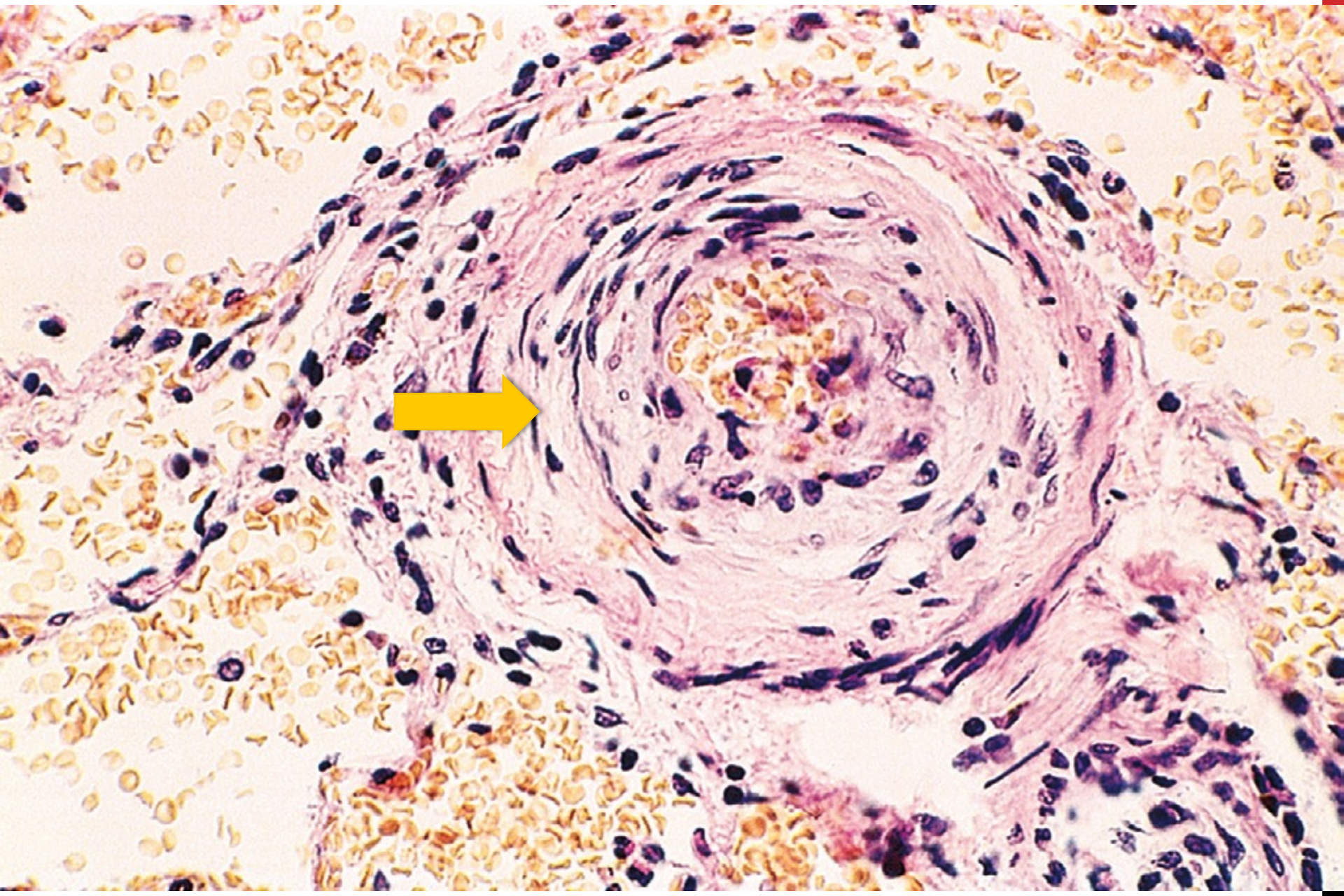
- **Pulmonary arterial hypertension:**
  - heritable forms of pulmonary hypertension
  - affects small pulmonary muscular arterioles
    - Examples: connective tissue diseases, human immunodeficiency virus, and congenital heart disease with left to right shunts

- **Pulmonary hypertension due to left-sided heart disease:**
    - including systolic and diastolic dysfunction and valvular disease

- **Pulmonary hypertension due to lung diseases and/or hypoxia**:
    - including COPD and interstitial lung disease
- **Chronic thromboembolic pulmonary hypertension**
- **Pulmonary hypertension with unclear or multifactorial mechanisms**

MORPHOLOGY:

- **Medial hypertrophy of the pulmonary muscular and elastic arteries**
    - small arteries and arterioles
- **Pulmonary arterial atherosclerosis**
    - pulmonary artery and its major branches
- **Right ventricular hypertrophy**



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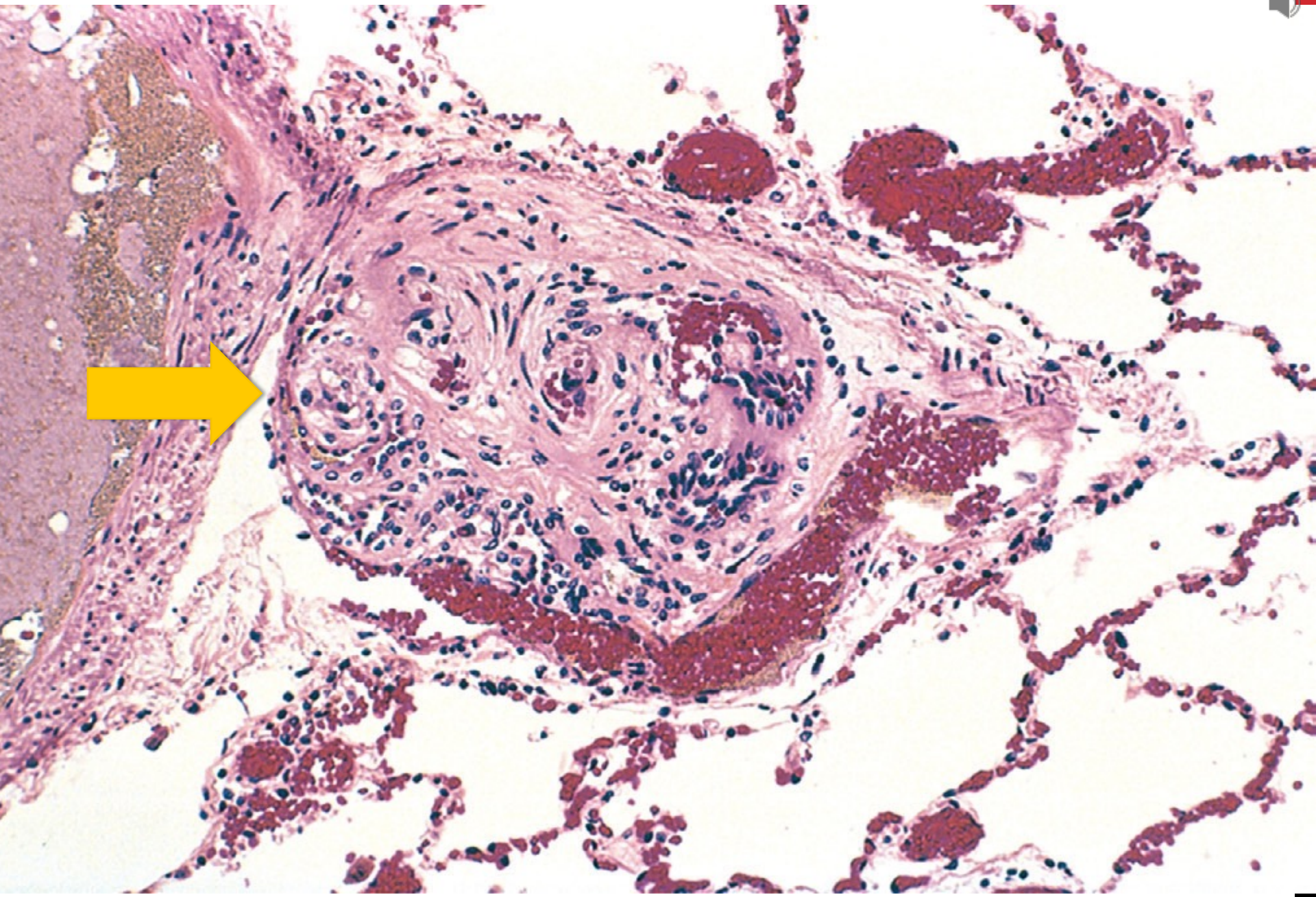
- **Plexiform lesion:**

  - uncommon

  - a tuft of capillary formations producing a network, or web, that spans the lumens of dilated thin-walled, small arteries and may extend outside the vessel.



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DIFFUSE ALVEOLAR HEMORRHAGE SYNDROMES

- Complication of some interstitial lung disorders.

Includes:

1. Goodpasture syndrome
2. Granulomatosis with polyangiitis
3. Idiopathic pulmonary hemosiderosis

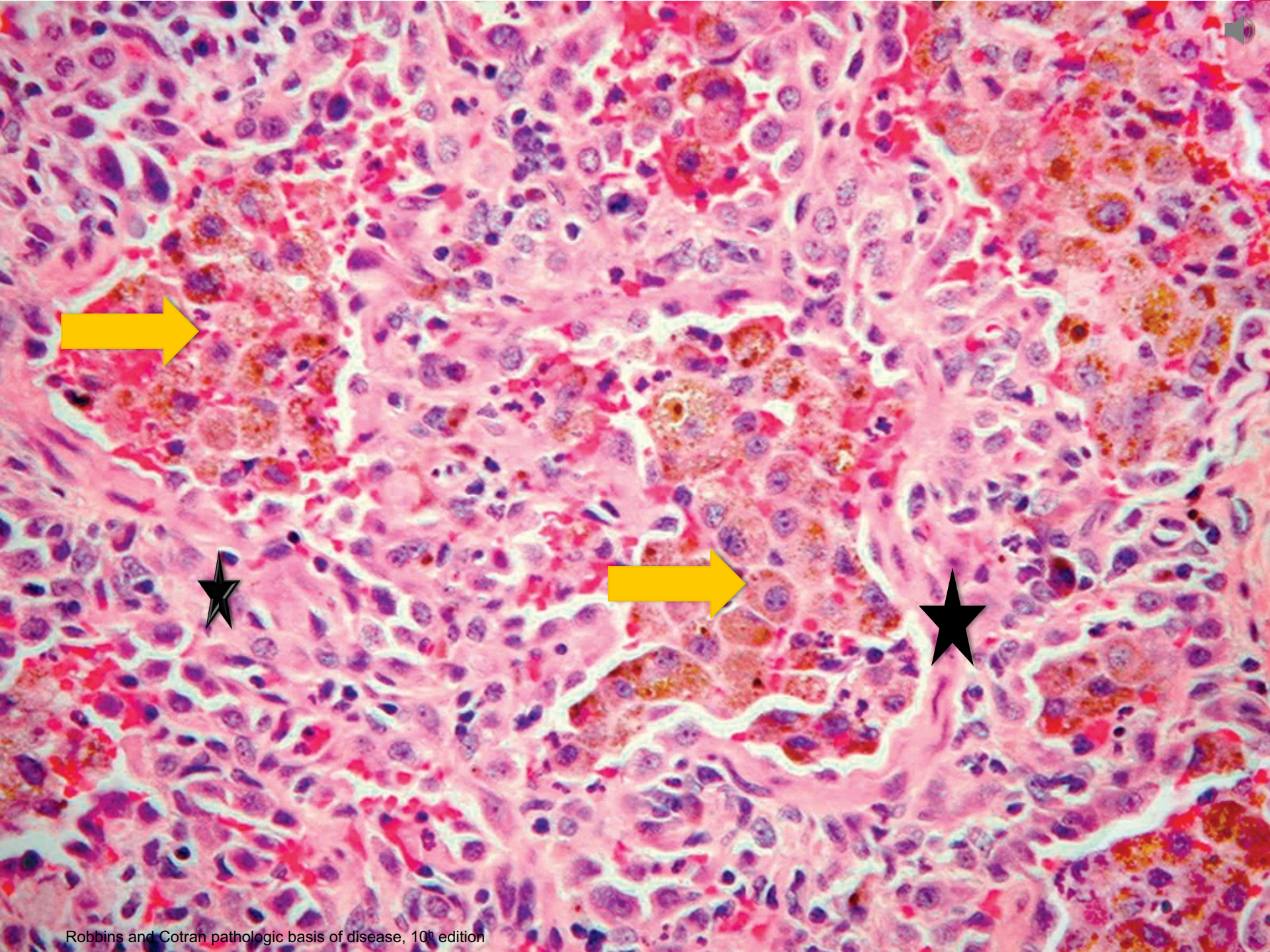
GOODPASTURE SYNDROME:

- **Is an uncommon autoimmune disease in which lung and kidney injury are caused by circulating autoantibodies against certain domains of type IV collagen.**
  - type IV collagen is intrinsic to the basement membranes of renal glomeruli and pulmonary alveoli
- **Results in necrotizing hemorrhagic interstitial pneumonitis and rapidly progressive glomerulonephritis.**

MORPHOLOGY:

- **Grossly,** red-brown consolidation due to **diffuse alveolar hemorrhage**

- **Microscopically:**
  - Focal necrosis of alveolar wall with intraalveolar hemorrhage,
  - Fibrous thickening of septa
  - Hypertrophic type II pneumocytes.
  - Abundant hemosiderin
  - Linear pattern of immunoglobulin deposition (IgG, sometimes IgA or IgM) seen along the alveolar septa.



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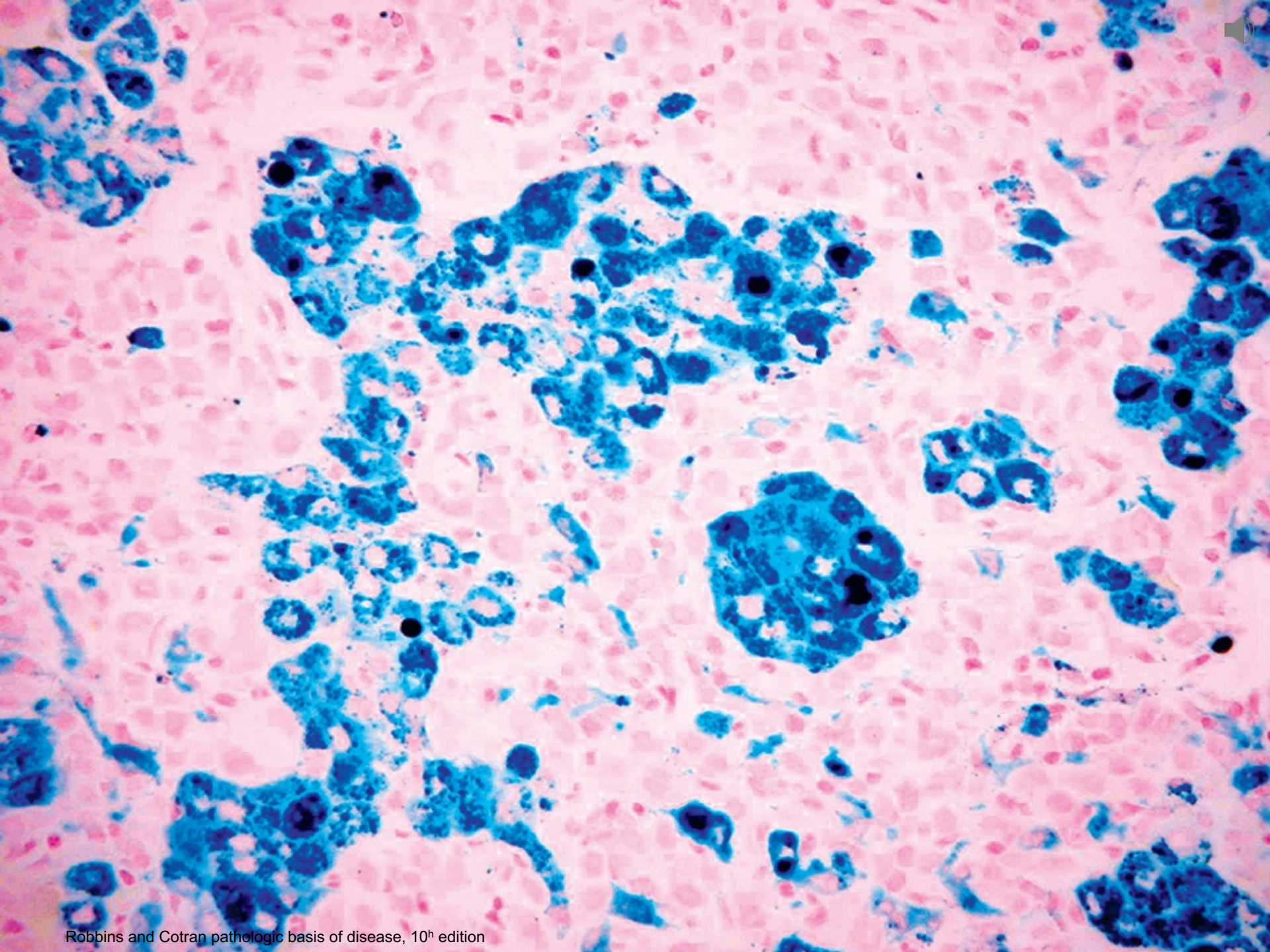

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CLINICAL FEATURES:

- Teens and twenties
- Males > females
- Active smokers
- Plasmapheresis and immunosuppressive therapy , renal transplantation

GRANULOMATOSIS AND POLYANGIITIS

- Formerly called **Wegener granulomatosis**
- >80% of patients develop upper-respiratory or pulmonary manifestations.
- The lung lesions are characterized by a combination of necrotizing vasculitis (“angiitis”) and parenchymal necrotizing granulomatous inflammation.

- The signs and symptoms of the upper-respiratory tract involvement (chronic sinusitis, epistaxis, nasal perforation) and the lungs (cough, hemoptysis, chest pain).

- Focal necrotizing, often crescentic, glomerulonephritis.

- Anti-neutrophil cytoplasmic antibodies (PR3- ANCAs) are present in close to 95% of cases

THANK YOU!