* **Pathology of pulmonary vascular disease**
* **Dr.Ashraf Abdelfatah Deyab**
* **Assistant Professor of Pathology**
* **Faculty of Medicine**
* **Almajma’ah University**
* **Pulmonary vascular disease**
* **Type of pulmonary circulation:**
* **Types of pulmonary vascular disease**
* **Objectives**
**Pathology of Pulmonary Vascular Diseases**
* To discuss the etiology**1**, morphological**2** features and clinical consequencesof P**ulmonary embolism(PE)**
* To describe the pathogenesis**1,**morphology**2** and clinical features**3** of P**ulmonary hypertension (PH).**
* **Pulmonary embolism (PE)**
* **Definition*:***
* **Impaction** of a **thrombus** or **foreign** matter in the pulmonary vascular bed as secondary of other conditions, leads to complications and death.
* **Process:**
* **Blood clots formation** 🡪**BREAK** & **TRAVEL** to **occlude** the **pulmonary arteries& branches** (one or more).
* T**ypes**: **(1) Thrombotic (2) Non-thrombotic**
* **Source of Non-thrombotic PE (rare):**
* **1. Tumors,**
* **2. Air bubbles. 3. Amniotic fluid.**
* **4. Fat.**
* **The venous thromboembolism (VTE) refers to DVT, PE, or to a combination of both.**
* **Rudolf Virchow** "Father of Pathology”

* (>90%) of PE cases are originating from the deep veins e.g popliteal vein.
* **All predisposing factors to DVT is well explained by him.**
* **Virchow-triad**
* **Stasis of blood flow.**
* **Endothelium Injury** (irritation, trauma )
* **Hypercoagulablity** (Thrombophilia).
* **PE Predisposing\Risk factors:**
* I**nherited** Hypercoagulable states, (AT III def., protein C, S deficiency).
* **Acquired**
* **Immobilization-** Bed rest
* **Post-operative** (Hip, legs, abdomen)
* Severe blood loss and trauma (fractures & burns)
* **Women** (Pregnant, oral contraceptive rich in ER)
* Varicose veins
* Advancing age.
* Obesity, smoking
* Malignancy
* DM
* Cardiac diseases-CHF, HTN, MI, Fibrillation
* 1ry polycythemia.
* Race
* **PE morphology**-
* Origin
* **1.Thrombotic** in origin- most common.
* 2. **Veins** > Arteries.
* 3. Typical sites: **Deep veins of the calf and Deep pelvic veins**
* **Large-vessel in situ thromboses** are rare.
* **PE morphology**
* may lodge in various sites in the pulmonary arterial tree.
* **1) Large emboli** lodge the in the main pulmonary artery or its major branches or at the bifurcation as **a saddle embolus (**sudden death).

* **PE morphology based on site and size**

**2) Hemorrhages at the periphery (small emboli).**

**3) Lung infarction- Wedge shaped,** (base at the pleural surface & the apex pointing to the hilus of the lung)- hemorrhagic.

* **4) Thrombus\clot can be distinguished from a post-mortem clot by the presence of the lines of Zahn in the thrombus.**

* **Microscopic of pulmonary infarct**
* **Ischemic necrosis of the lung within the area of hemorrhages, alveolar, bronchioles, BV**
* **Cellular events with Hemosiderin deposits**

**3) Infected embolus, reveals intense neutrophilic inflammatory reaction referred as septic infarcts= abscesses.**

**4) Fibrous replacement – converts into a contracted scar.**

* **PE morphology- based on emboli source**

* **PE- Clinical course**
* 60-80% are clinically silent.
* 5% sudden death (large emboli).
* < 3% of cases recurrent pulmonary infarcts, result **in pulmonary HTN& RIGHT HF.**
* **Common symptoms& signs**
* **Diagnosis: D-DIMER, USG-DOPPLER, CT, MRI**
* **PE - The clinical effect& Consequences**

**The clinical effect depends on**

 **Two main pathophysiologic “effects” consequences:**

* PE - outcome

 **1) Occlusion** of a major vessels leads to:

 I. **Sudden DEATH** 🡪 (Unresolved + complication, HF)

* **2) Occlusion** of a smaller vessel:

I- **No effect** if the bronchial circulation is good. (resolved)

II - **Pulmonary HT 🡪 (**If small and multiple +Recurrence).

III. **Pulmonary infarction**

* **PULMONARY HYPERTENSION (PH)
objectives**
* To describe the pathogenesis, morphology and clinical features of **pulmonary hypertension (PH).**
* WHAT IS PULMONARY HYPERTENSION
* **Definition:**
* is Hemodynamic **SERIOUS& FATAL** illness chr. by **high BP** in the affected **BV** in the lungs and **Right side** of the heart- **due to narrowed, blocked or destroyed BV.**
* **BV= Pulmonary Arteries, capillaries& veins.**
* The mean pulmonary artery pressure (mPAP) reaches BP> 25 mm Hg at rest & > 30 mm Hg during exercise.(measured by right heart catheterization).
* **PH-** isn't curable, treatments are available that can help lessen symptoms and improve quality of life.
* **Complications🡪 RHF, CLOT, BLEEDING (hemoptysis)**, **Arrhythmia**
* **What’s the main causes of PH?**

The pressure in the lung BV increased for two reasons:

* **1) Increased blood flow.**
* **2) Increased resistance within the pulmonary circulation. (narrowed, destroyed, blocked)**

Can be classified into three main causes based on etiology:

* **Secondary pulmonary hypertension:**
* **caused by another medical problem, e.g.**
* **PE, CT disease, Sickle cell anemia**
* **COPD, Lung fibrosis& scarring, HIV, Drugs-induced.**
* **Cardiac diseases, LHF, vasculitis.**
* **2- Primary pulmonary hypertension (Familial)**

 - Rare, **Mutations, autosomal dominant inheritance**.

 - No underlying cause. Patients are rather sensitive to any vasoconstrictors.

* **3) Idiopathic PH:**
* Sporadic, requires exclusion of others.
* Usually women 20-40 years old, some time children.
* **PH- Pathogenesis**
* **Occurs in Primary PH(familial)🡪 M**utations in the bone morphogenetic protein receptor type 2*(****BMPR2****)****🡪*** *BV thickening & occulsion.*
* **Occurs in Secondary PH 🡪 produced endothelial cell dysfunction e.g.- Leftto-right shunts (Mechanical),Thrombo-embolism, (biochemical injury produced by fibrin**).
* **Occurs in Secondary PH 🡪 Platelet Aggregation& adhesion+ Endothelial activation+ Cytokines production + vasospastic effect.**

* PH morphology

**1. Medial hypertrophy**

**2., Atheromatous deposits**.

**3.Initimal fibrosis-narrowing**

 **4. Organizing or recanalized thrombi**, with **coexistence of diffuse fibrosis** this **favors recurrence.**

* **5. Alveolar hemorrhages**
* Morphology of PH-Gross changes

**Pulmonary hypertension, reveal atheroma formation, usually limited to large vessels**

**6- Plexiform lesion-**in small arteriesmultichannel **.**

- Associated with :

* **Idiopathic& primary PH+**
* **Congenital heart disease with left-to-right shunts.**
* **PH Clinical features**
* **Sign& symptoms:**
* Like HTN are **subtle** in the early stages**.**
* **Hidden** by underlying diseases.
* **Varying** from pt. to pt.
* **Initial Symptoms:** dyspnea, cough, fatigue, chest angina-like pain, slowed growth (in child).
* **Overtime Severe respiratory distress**, **cyanosis**, and **right ventricular hypertrophy, RHF.**
* **PH outcome:**  **Death** from decompensated cor pulmonale, often with superimposed thromboembolism and pneumonia.

THE END