Pulmonary Vascular Disorders 2023

Pulmonary Hypertension
Cor-Pulmonale
Pulmonary Embolism
Venous Air Embolism

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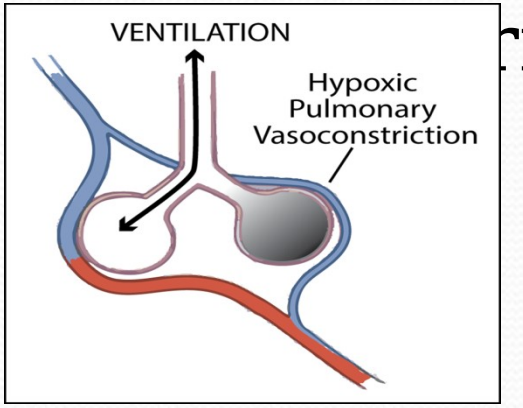
Pulmonary System

- Lungs need to remain perfused to support other organ systems
- *Systemic circulation* ***dilates*** in response to hypoxia, *pulmonary vasculature* ***constricts***
- Pulmonary vasoconstriction occurs *locally* in response to hypoxia
- In respiratory failure as SaO_2 falls, pulmonary arteri rises



Pulmonary Hypertension

- Can be Primary (idiopathic)

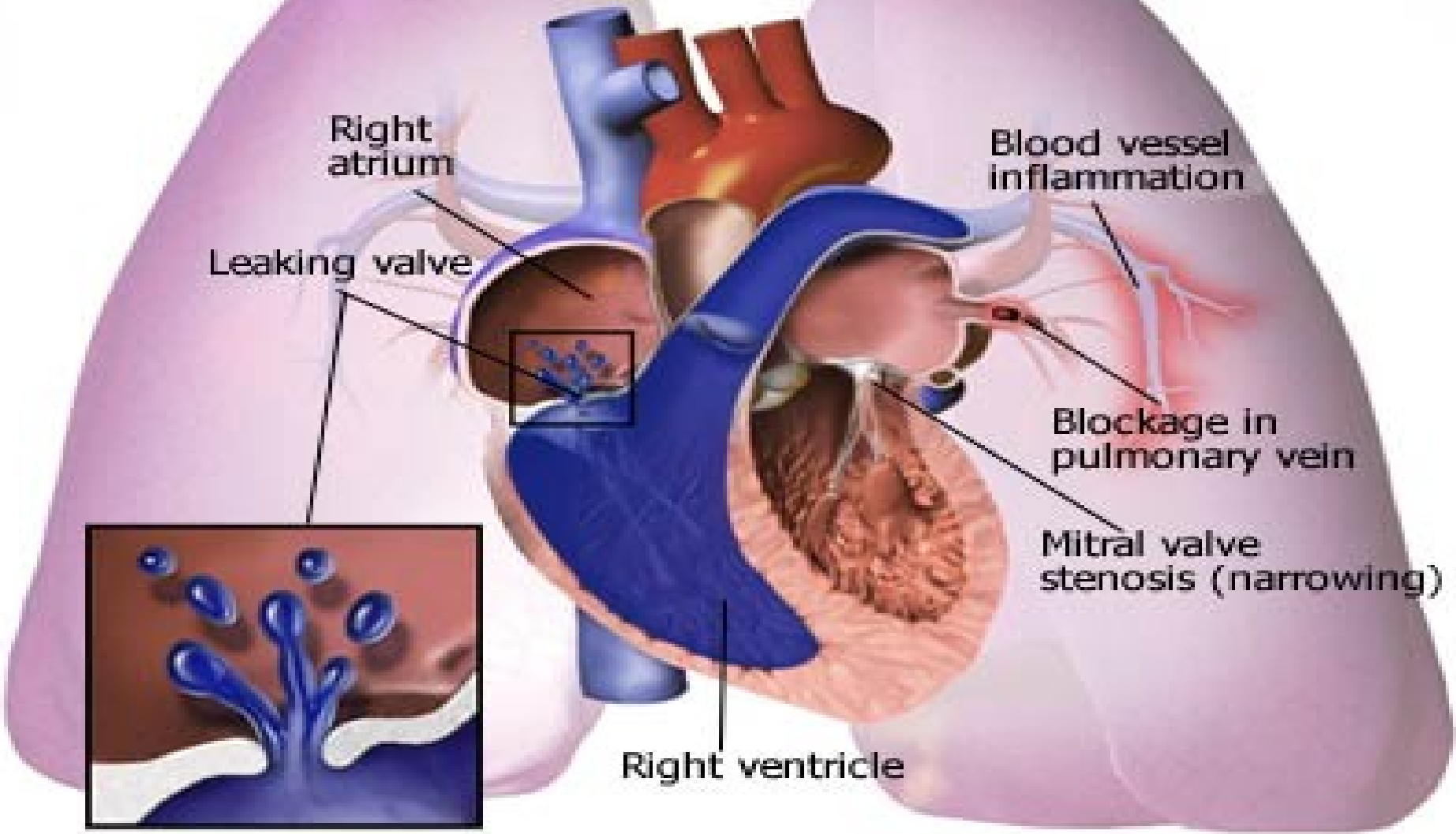
  or Secondary – result of other diseases, often cardiac)
- Elevated pulmonary pressure results from increase in pulmonary vascular resistance to blood flow through small arteries/ arterioles
- Normal mean PAP = 12-16
- **Pulm HTN** = Mean artery pressure > 25mm Hg (at rest) or > 30 mm Hg (while exercising)

Pulmonary Hypertension

- Increasing hypoxia → increased pulmonary hypertension
  - May have 50% of area of normal pulmonary bed compromised due to hypoxia in pulmonary hypertension before symptoms occur
- Reduced size of pulmonary vascular bed- causes ↑ pressure with circulation of blood
- Death due to right ventricular failure, pulmonary embolism or hemorrhage/ sudden death

Pulmonary Hypertension

Can be caused by a number of factors, all of which force the heart's right side to work harder to pump blood to the lungs. The right chambers may enlarge as they struggle to function, and blood is often forced backward through the tricuspid valve.



Primary pulmonary hypertension (rare)

- Persistent elevated pulmonary pressures
- Cause: Deficient release of vasodilator mediators from the pulmonary epithelium
- May be genetic
- **Unknown etiology (idiopathic)**

Primary Pulmonary Hypertension

- Women/Men 4:1 average; 36 ave. age
- Symptoms: dyspnea(60%), weakness (19%), recurrent syncope (13%)

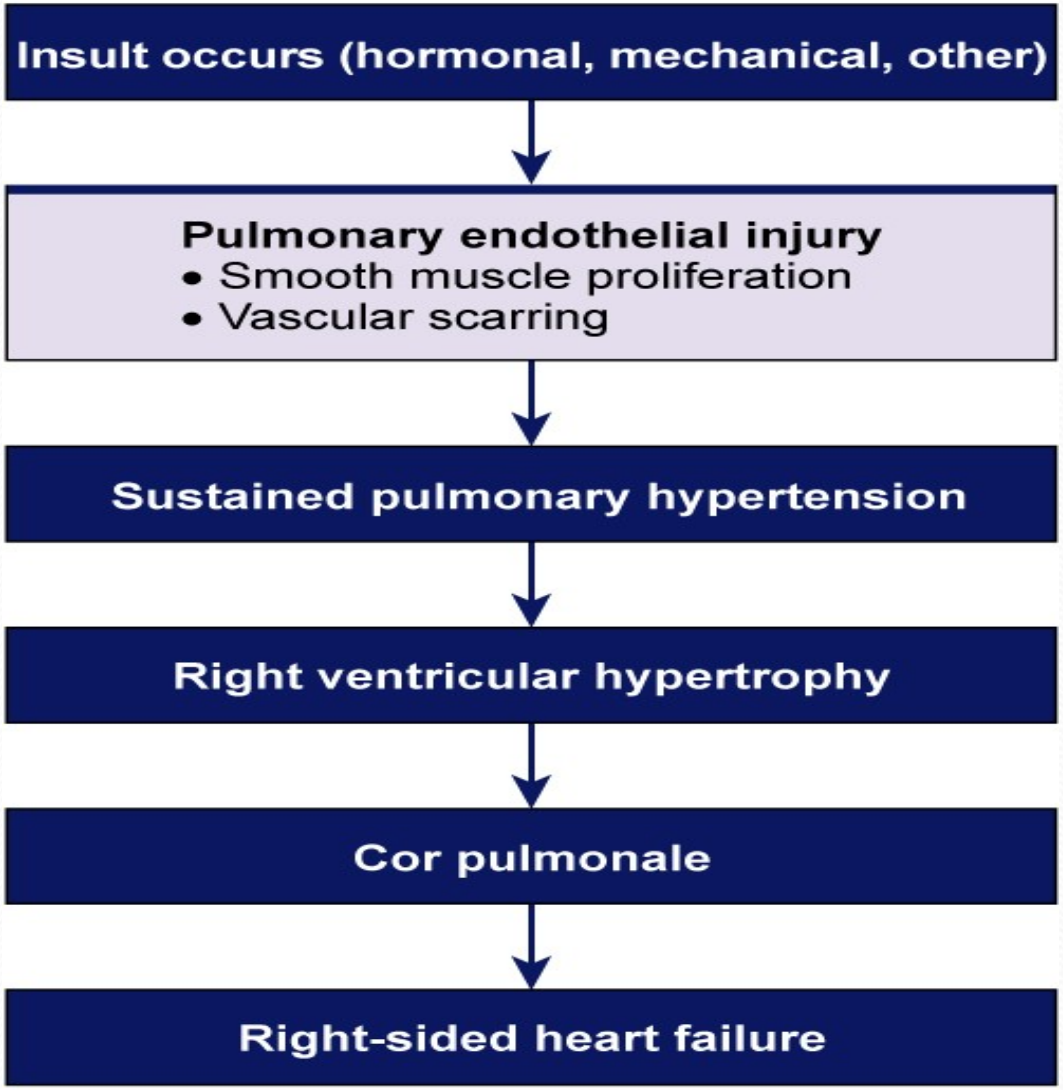
Diagnostic studies

- CXR - may show enlarged pulmonary artery, clear lungs, cardiomegaly, enlarged R heart
- Chest CT - enlarged pulmonary artery and vasculature changes

Primary Pulmonary Hypertension

- ABGs- initially decreased $PaCO_2$ -normal or low PaO_2
- PFTs-restrictive lung volume pattern on PFTs or a normal pattern
- ECG/ECHO – RV hypertrophy and other changes
- **Rt-sided Cardiac Catheterization**
  - – measure PAP, CO, LVFP

PATHOPHYSIOLOGY MAP



Treatment of PPH

- *Early recognition essential to prevent progression:* **No cure -** treat symptoms
- Oxygen- given to treat hypoxemia - goal of >/= 90% saturation; prevent pulmonary vasoconstriction
- Anticoagulants- prevent thrombus formation; coumadin - INR in 2 - 3 range
- Diuretics- reduce dyspnea & peripheral edema; may reduce volume overload of right sided failure (overuse causes decrease in preload/cardiac output- must monitor electrolytes)

Treatment of PPH

- Calcium channel blockers (Oral)
  - Vasodilator, reduces afterload & improves right sided failure. Reduction of PAP in higher doses
  - Diltiazem (Cardizem) , Nifedipine (Procardia)
- Phosphodiesterase inhibitors (Oral)
  - Sildenafil (Viagra), Milrinone
  - Watch for orthostatic hypotension
- Vasodilators (IV)
  - Prostacyclin IV-promotes pulmonary vasodilation/ reduces pulmonary vascular resistance
    - no effect on SVR, platelet aggregation inhibitor **(6 minute half-life)**
    - Can see rapid deterioration in abrupt withdrawal
- Vasodilators (Inhaled)
  - Prostacyclin inhalant, 6-9 X / day
- Lung transplant
  - Limited availability
  - May still re-occur

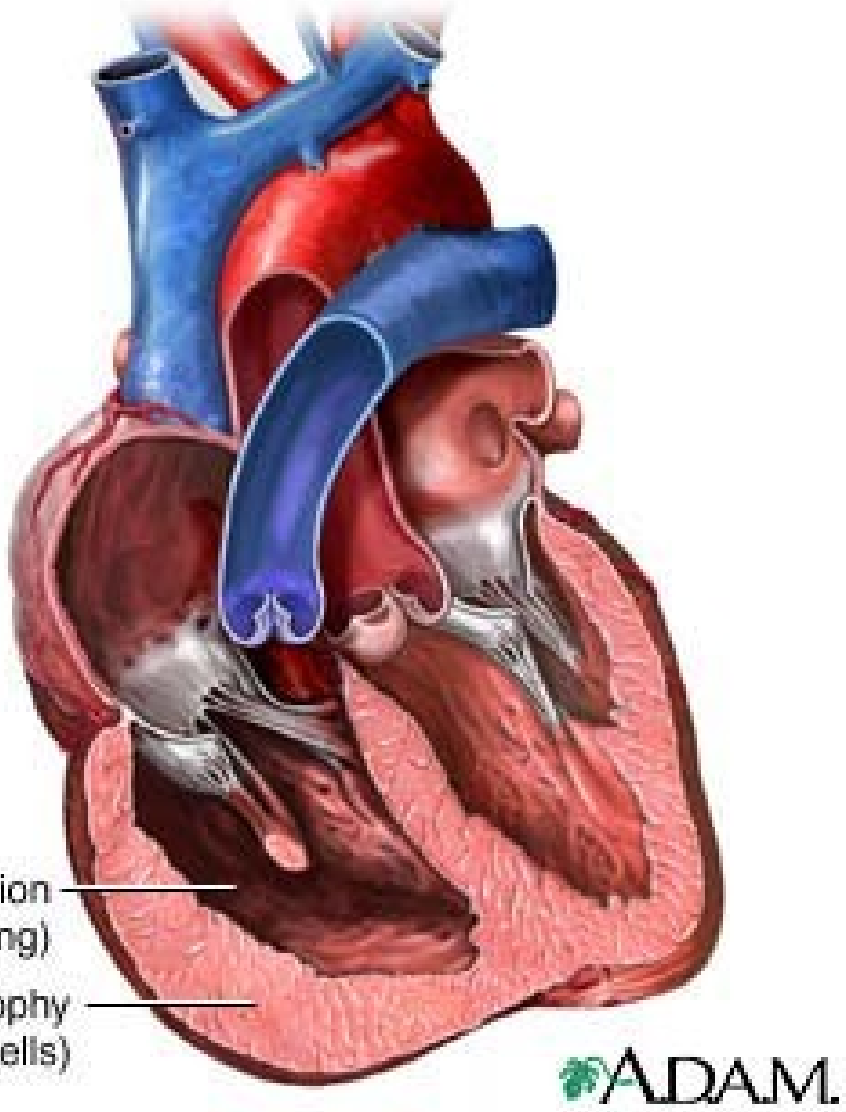
Secondary Pulmonary Hypertension

- Occurs when another primary disease *causes* increase in pulmonary artery pressures – COPD # 1
- Symptoms reflect underlying disease *causing ↑ PVR – 3 reasons*
  - *1. Capillary loss (COPD)*
  - *2. Stiffening of pulmonary vasculature (Pulm. Fibrosis)*
  - *3. Obst. Blood flow (PE)*
- Diagnostic studies (see PPH)
- Treatments
  - treat underlying primary disorder
  - initiate PPH therapies to manage symptoms

Cor Pulmonale

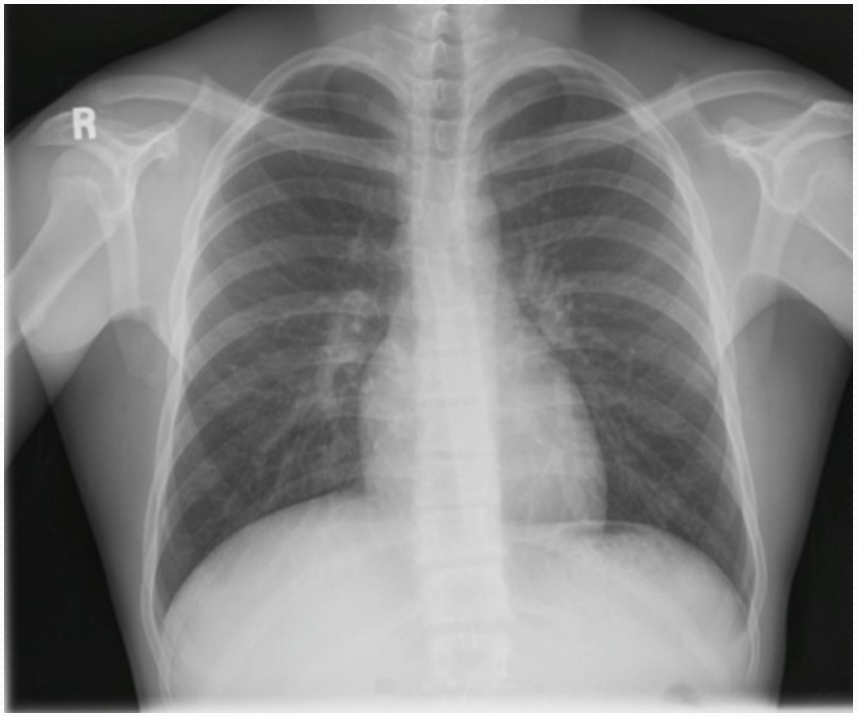
- **Right ventricular enlargement**
- Condition of hypertrophy/dilation of right ventricle of heart resulting from disease process that affects function or structure of lung or its vasculature
- Most common cause is **COPD**
- Acute cor pulmonale
  - associated with pulmonary embolism causes RV *dilation*
- Chronic cor pulmonale
  - associated with/caused by various disorders & characterized by RV *hypertrophy*

Cor pulmonale, or right-sided heart failure, is an enlargement of the right ventricle due to high blood pressure in the lungs usually caused by chronic lung disease

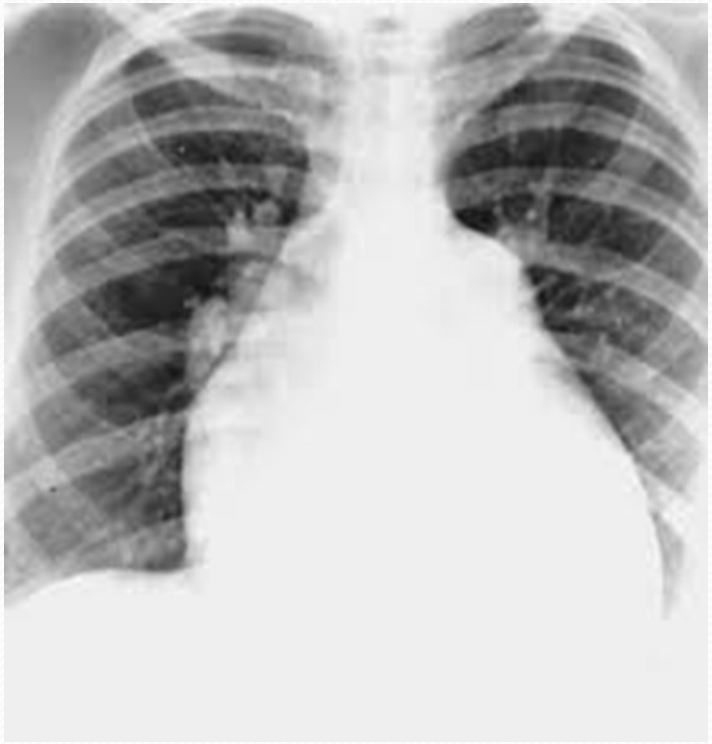


X ray comparison

Normal Chest X ray



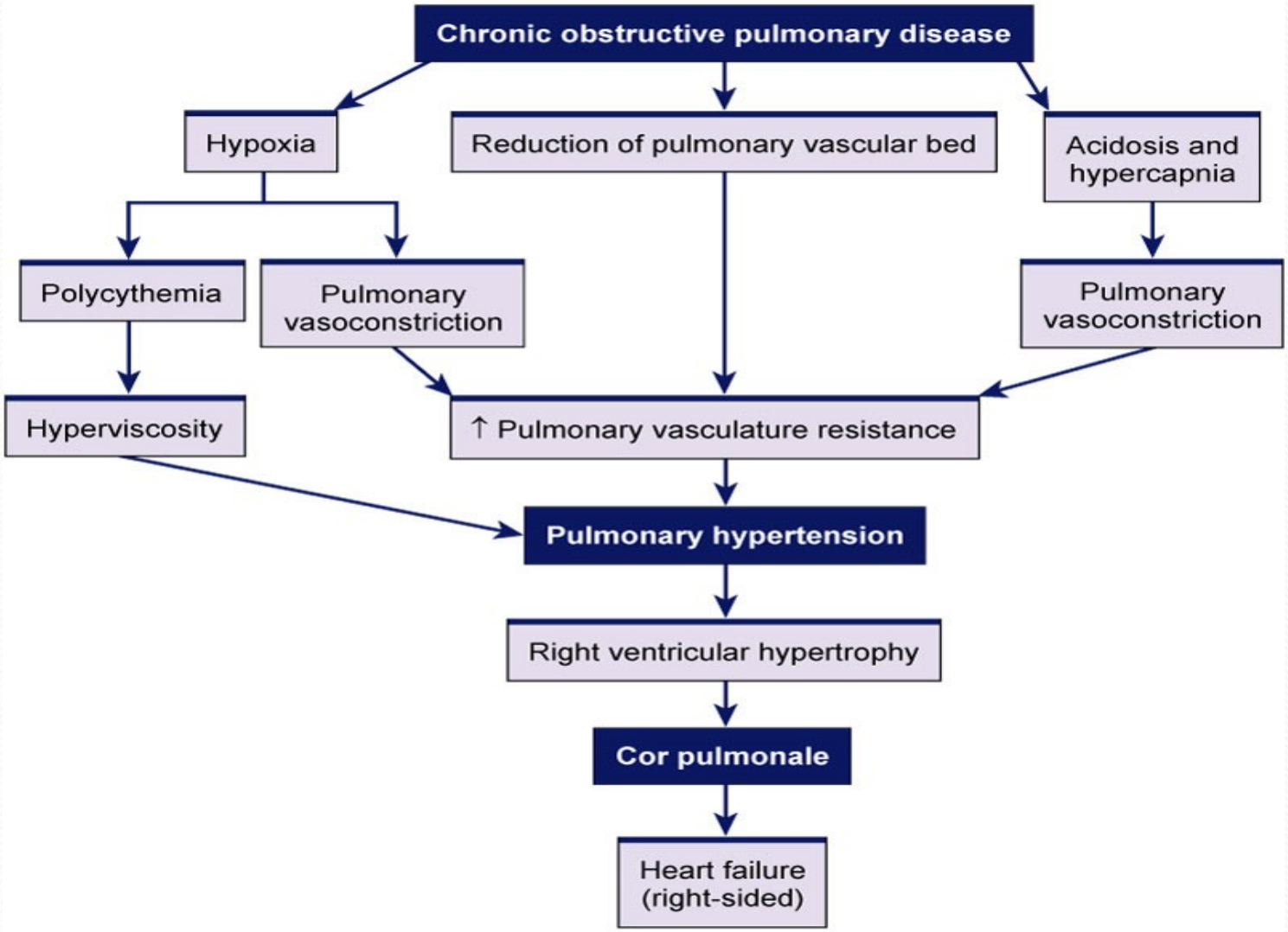
Cor Pulmonale Chest X ray



Cor Pulmonale

- RV doesn’t have full compensating ability (unlike LV) dilates-fails in acute situations
- Complication of PE & Pulmonary infarct
- Normally, RV doesn't work hard -pulmonary resistance low - Right heart flow passes with little problem
- RV failure leads to increased pressure-causing RV to bulge into intraventricular septum - reducing left ventricular volume

PATHOPHYSIOLOGY MAP



Cor Pulmonale: Clinical Manifestations & Diagnostic Studies

1. Dyspnea
2. Lung sounds normal or crackles at bases
3. Distended neck veins, peripheral edema (+2), bounding pulses, fatigue, hepatomegaly with RUQ tenderness, weight gain
4. Chest xray-enlarged right ventricle/pulmonary artery
5. BNP elevated
6. ABG's
7. Cardiac Cath

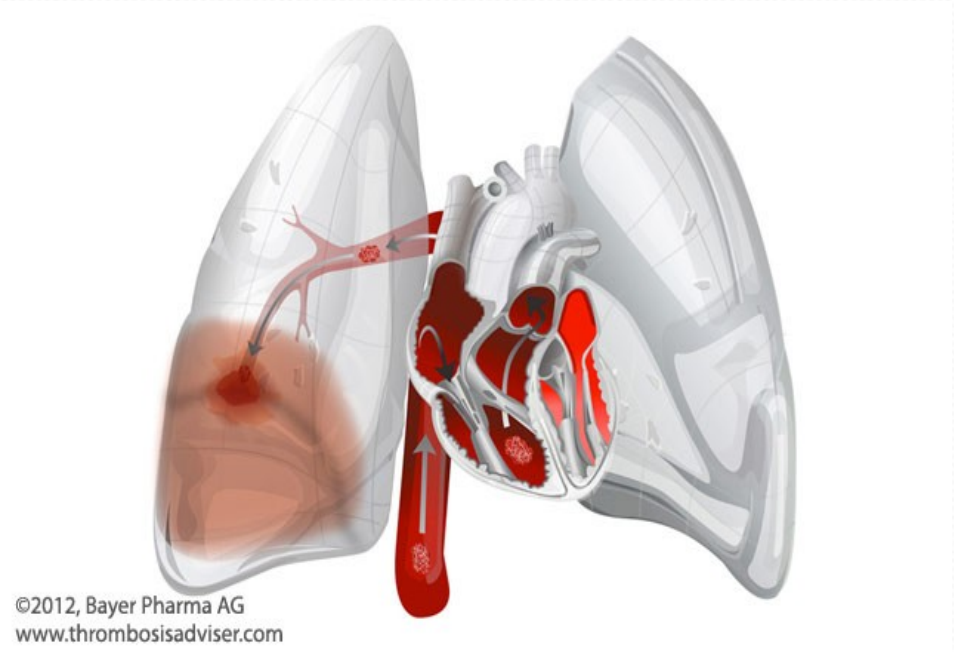
Cor Pulmonale

Important to treat symptoms

1. Oxygen therapy/low flow
2. Bronchodilators / Theophylline
3. Diuretics (electrolyte issues)
4. Low-sodium diet/fluid restriction/frequent & small meals
5. Digoxin
6. Vasodilators
7. Calcium channel blockers
8. Anticoagulants
9. ACE inhibitors

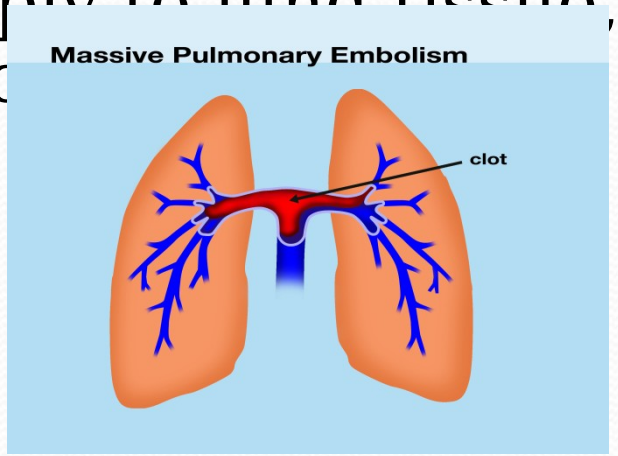
Pulmonary Embolism

- 3rd leading cause of death in US hospitalized patients
- 50,000 – 100,000 deaths/yr
- Occurs in 1 per 1000
- 44% confirmed DVT



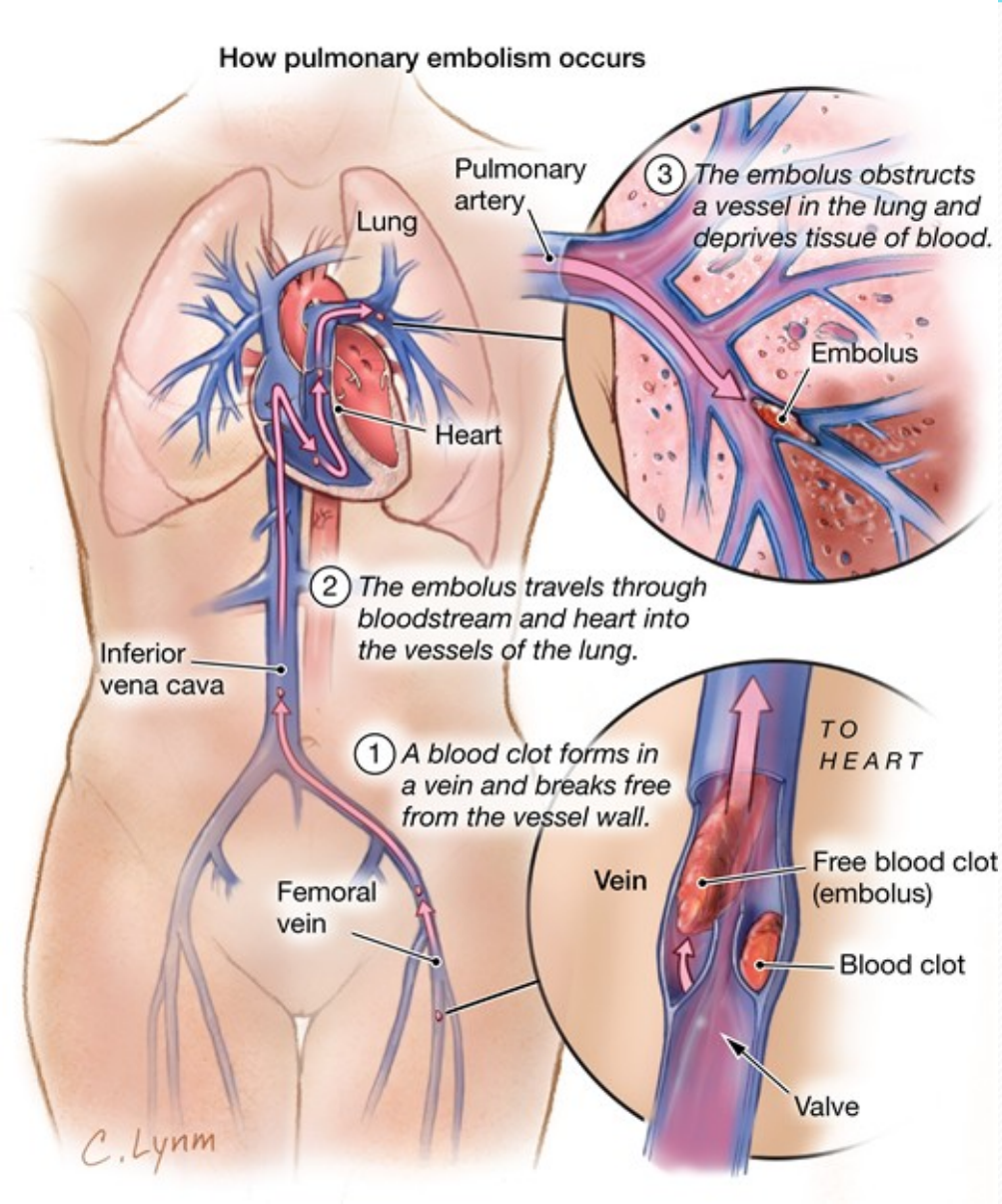
Pulmonary Embolism

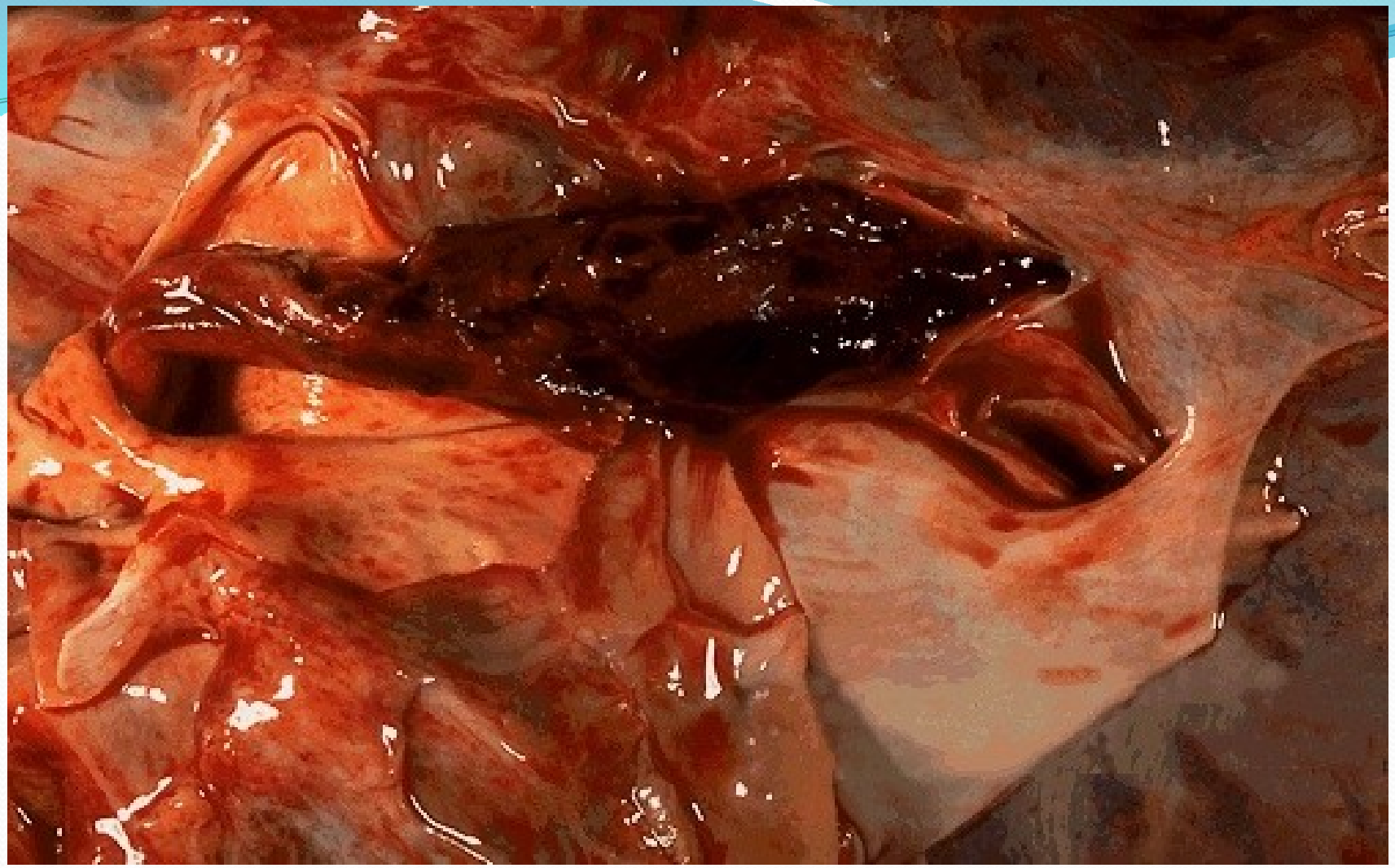
- Pulmonary embolism
  - blockage of pulmonary artery or one of its branches by foreign matter, thrombus
  - from peripheral vein, air, tumor tissue, amniotic fluid, bone marrow, foreign IV material, bacterial vegetations, right side of heart (thromboembolism from atrial fibrillation).
- Blockage obstructs blood supply to lung tissue, ↑ RV workload, ↓ perfusion to



Pulmonary Embolism

- Most common cause: Deep Vein Thrombosis (DVT)
  - Over 90% of all cases are from DVT's
  - VTE = venous thromboembolism (DVT→PE)
  - Greater risk of above knee clots (Iliofemoral vein) embolizing than below knee clots
- Predisposed to developing DVTs: Virchow's triad:
  - 1. Venous stasis
  - 2. Endothelial injury (intimal)
  - 3. Hypercoagulability e.g., deficiency in antithrombin III, protein C - protein S, dehydration


How pulmonary embolism occurs
Pulmonary artery
Lung
3 The embolus obstructs a vessel in the lung and deprives tissue of blood.
Embolus
Heart
2 The embolus travels through bloodstream and heart into the vessels of the lung.
Inferior vena cava
1 A blood clot forms in a vein and breaks free from the vessel wall.
TO HEART
Vein
Free blood clot (embolus)
Femoral vein
Blood clot
Valve
C.Lynm



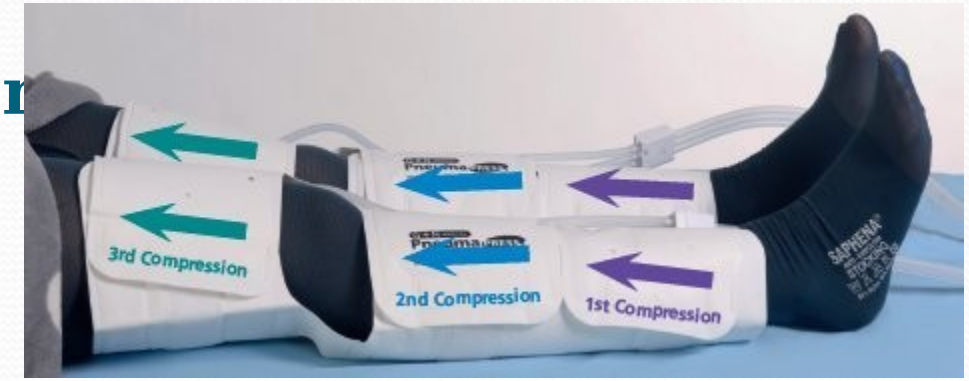
Pulmonary trunk /pulmonary arteries to right/left lungs opened to reveal a large "saddle" pulmonary thromboembolus. Such an embolus will kill.

Clinical States at increased risk for DVTs

1. **Immobility,** i.e. Spinal cord injury, stroke, prolonged travel, especially flights, bedrest
2. Recent surgery (w/in past 3 months)
3. Prior DVT
4. A-fib (Rt side of heart)
5. Venous insufficiency, smoking
6. Recent central venous instrumentation (CL, PICC)
7. Pregnancy (especially post partum), birth control
8. Malignant neoplasms
9. Clotting disorders

Prevention of DVT:

- low-dose heparin gtt (in low-moderate risk pts)
- warfarin (in high risk pop)
- Lovenox (Enoxaparin)
- Aspirin therapy
- EPC; best to use anticoagulants and compression together
- **Early ambulation**



Clinical Manifestations

Symptoms may begin slowly or suddenly:

1. Sudden onset dyspnea
2. Feelings of apprehension or anxiety, doom
3. Tachypnea
4. Tachycardia
5. Mild – moderate hypoxemia
6. Low $PaCO_2$
7. Hemoptysis

Massive PE

- Shock
  - Pallor
  - Severe dyspnea
  - Crushing chest pain
  - Pulse rapid and weak or PEA
  - Hypotension
- 30-60% mortality rate

Diagnostic Tests

1. Chest Xray – not diagnostic for PE!
2. **Spiral Chest CT, (CTA)** – reveals pulmonary abnormalities (most frequently used)
   - Caution: contrast allergies or poor renal clearance
3. V/Q Lung Scans - Perfusion Lung Scan
   - Effective, use serum albumin & isotope- if isotope does not show on Scan there is deficient perfusion [clot].
   - Ventilation Lung Scan- breathe in radioactive gas – area of decreased ventilation is area of embolus.

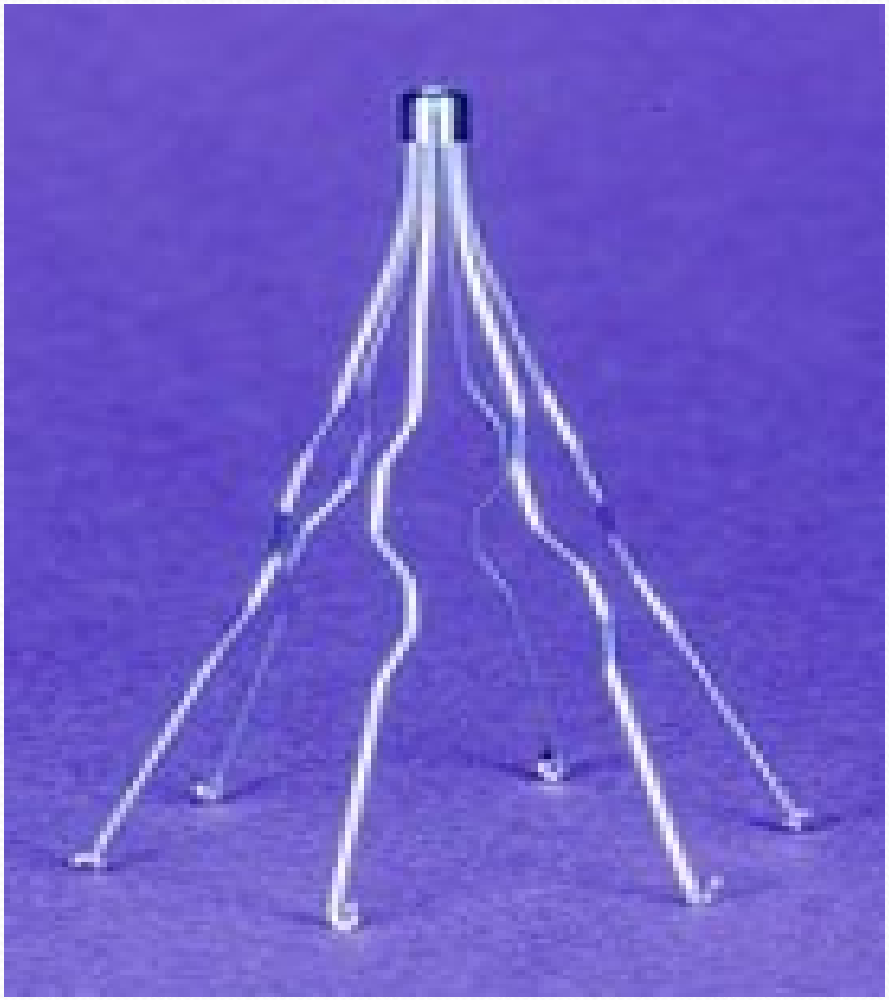
- Pulmonary Angiography-most specific but also most risk. Uses invasive cath & dye (expensive, seldom used now)

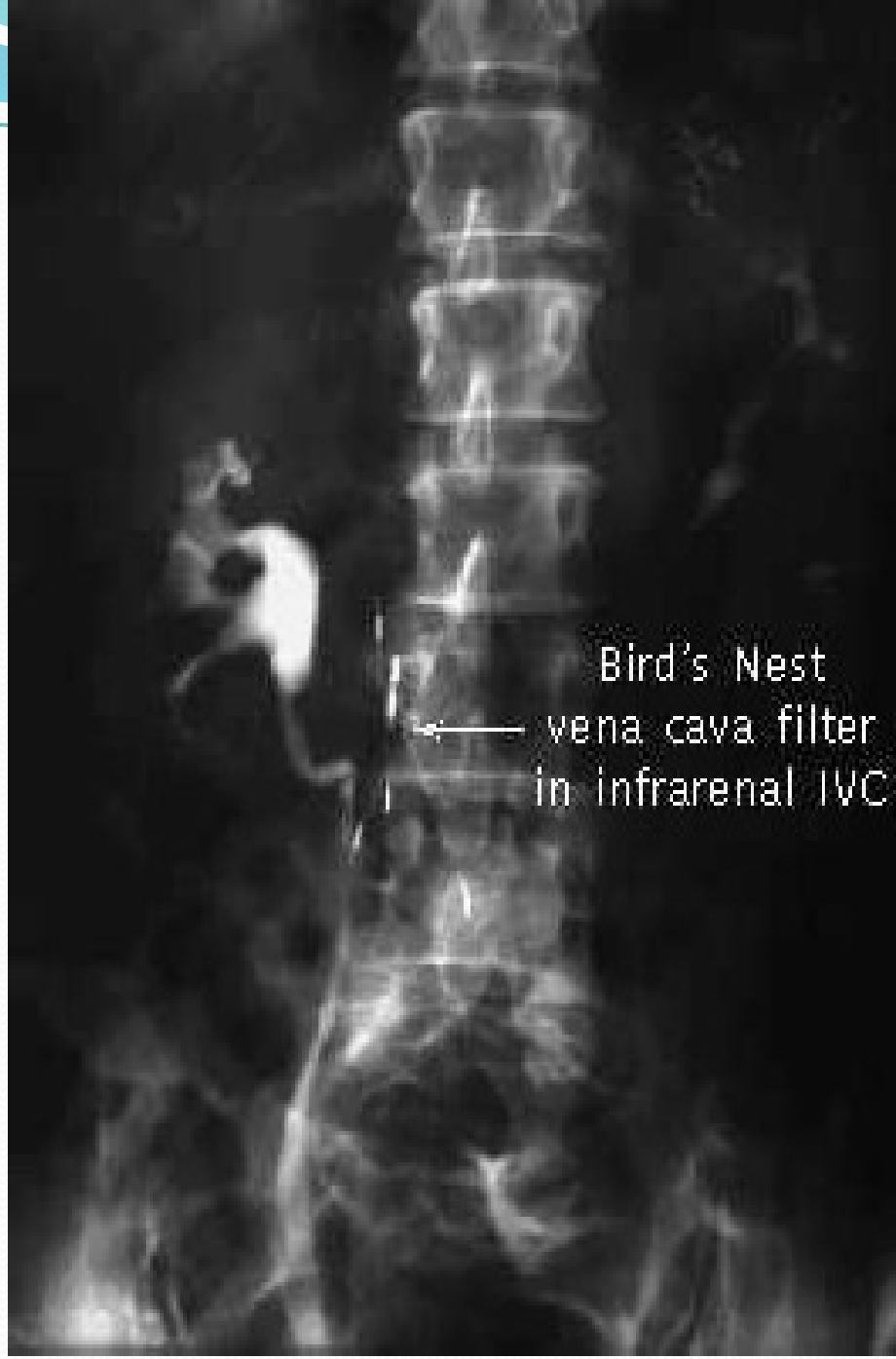
Diagnostic Tests

5. ABG's -PaO_2<80 mmHg, why? ; $PaCO_2$ < 35mmHg, why?
6. Doppler ultrasound (DVT)
7. D-dimer- produced during degradation of fibrin clot by plasmin
   - A "normal" D-dimer can r/o PE
8. ECG - not diagnostic

Collaborative Therapy

- Oxygen
- Thrombolytic agent – Alteplase, TPA
- IV Heparin or low molecular wt. heparin (Lovenox)
- Warfarin (coumadin) for long term
- Bedrest - for the first 2 or 3 days
- Narcotics for pain relief
- Inferior vena cava filters {Greenfield, Gunther Tulip, others}
- Pulmonary embolectomy in life-threatening situations (tricky procedure, can have 50% mortality rate)
- Monitoring aPTT, anti Xa, PT, INR, platelets, ABG's
- Assessments: VS, rhythm, lung sounds, LOC




Bird's Nest
vena cava filter
in infrarenal IVC

Complications of Clots

- Hypoxemia
- Hyperventilation
- Pulm. infarcts at site of injury → necrosis
- Pulm. HTN
- Right heart failure
- Hemorrhage
- Pleural effusion

Venous Air Embolism

- Venous air embolism (VAE) entry of air into venous system as result of trauma or iatrogenic complications
  - Chest trauma – Gun shot, stabbing
  - Misuse of IV catheters, central line access
- Effects on pulmonary vasculature lead to inflammatory changes in pulmonary vessels:
  - direct endothelial damage
  - accumulation of platelets, fibrin, neutrophils, and lipid droplets
- VAE results in right ventricular dysfunction and pulmonary injury

VAE

- Usually, small amounts of air do not produce symptoms because air is broken up in the capillary bed & absorbed from the circulation
- Estimated > 5 mL/kg causes acute right ventricular outflow obstruction, results in cardiogenic shock/circulatory arrest
- 2-3 mL air into cerebral circulation can be fatal

Causes: VAE

- Open chest trauma, blunt chest or abdominal trauma, neck or craniofacial injury injuries
- Procedures-CV (internal jugular or subclavian) pressure infusion of fluids/blood; thoracentesis, HD
- Accidents at home during use & care of long-term central catheters (eg, Broviac, Hickman, Port-A-Cath) lead to VAE
- Complication of orthopedic, neurosurgical, or cardiovascular procedures.
- VAE component of decompression injury

Clinical Manifestations: VAE

- Develop immediately following embolization similar to pulmonary thromboembolism
- Severity of symptoms related to degree of air entry/include:
  - Dyspnea, Chest pain
  - Tachycardia, Hypotension
  - Altered sensorium
  - Circulatory shock or sudden death (patients with severe VAE)

Treatment

- Administer 100% oxygen
- Intubate for significant respiratory distress or refractory hypoxemia
- Place patient in Trendelenburg position-
  - rotate toward left lateral decubitus position
  - Traps air in apex of ventricle, prevents ejection into pulmonary arterial system
  - maintains right ventricular output

Treatment

- If central line procedure in progress- immediately terminate & clamp line
- If CV catheter already present, aspirate from distal port to attempt to remove air
  - Catheter may have to be advanced for this to be successful
- For decompression injury - hyperbaric oxygen therapy

Deterrence/Prevention

- Avoid opening catheter during subsequent manipulation
- Valsalva maneuver during removal of CVC
- VAE can occur following catheter removal/may be delayed for 30 minutes or more; initial dressing after removal should be occlusive.
- Take particular care with known right-to-left shunts, since paradoxical embolism to arterial system can occur.
  - Arterial air embolism can cause organ damage