Imaging Approach to Pulmonary Hypertension

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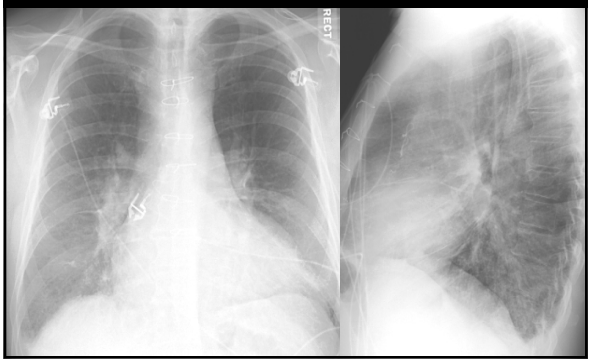
Disclosures

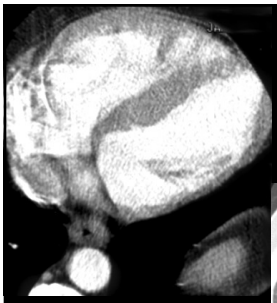
- No relevant disclosures for this talk
- Acknowledgment to Dr. Gopal Allada for some of the slides and clinical information

Lecture Objectives

- Briefly review classification, evaluation and therapies of pulmonary hypertension (PH).
- Review the radiographic approach.
- Role of multidetector CT in the evaluation of PAH and its classification.
- Implication of altered contrast flow dynamics.
- Understand that reports should address if a *post capillary etiology* is supected.

Introduction Patient: 67 Year Old Male with Progressive Dyspnea and Recurrent CHF Episodes

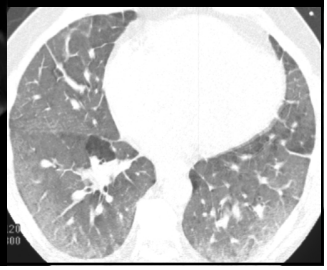




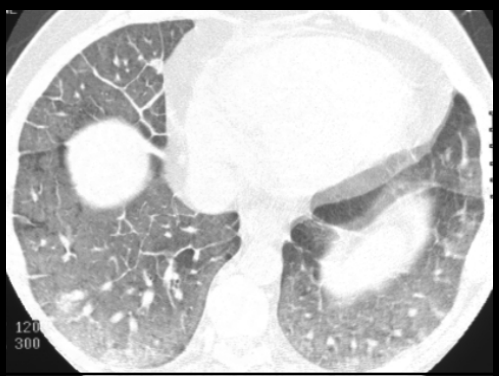
Septal Thickening and ground glass opacities. Looks like Hydrostatic Edema…

Moderate Pulmonary Hypertension with hypertrophy of RV wall.

Normal Left Ventricle Function and Pressures



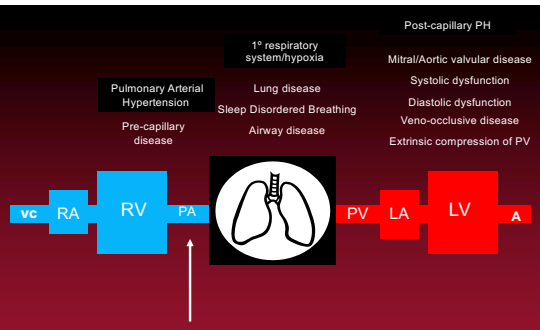
Possible Diagnosis? Report Impression?



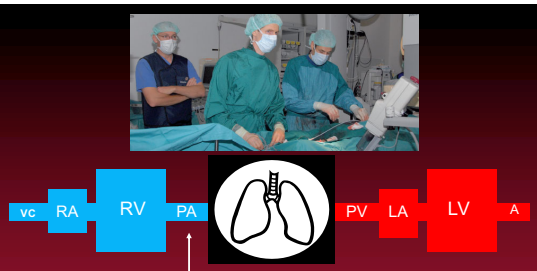
Pulmonary Arterial HTN: Definition

PAH Severity		Normal
Mild 25-40	Mean PAP ≥ 25 at rest or ≥ 30 mm Hg with exercise	PAP 12-16
Moderate 41-55	with	PCOP 5-12
Severe > 55	Mean PCOP/LVEDP ≤ 15 mm Hg*	PVR 1.5 – 2.5
	and	
	PVR > 3 Wood Units	

Farber H W, Loscalzo J. *N Engl J Med.* 2004;351:1655-1665.



Pulmonary Hypertension = abnormally high pressure in the pulmonary arteries



Pulmonary Arterial Hypertension = abnormally high pressure confined to the ***pre-capillary vessels***

Pulmonary Hypertension

Anatomic Organization

1. Pre-capillary
2. Capillary
3. Post-capillary

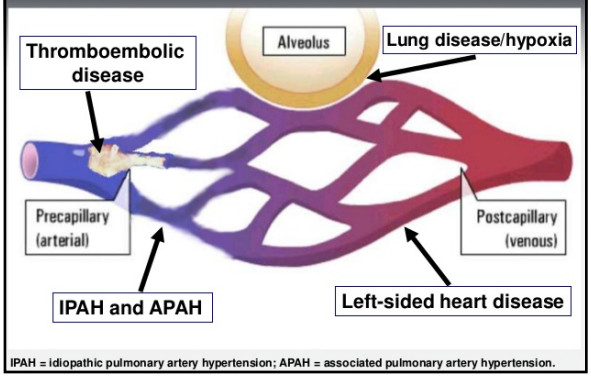
Physiological Organization

1. Increased flow
2. Chronic hypoxia
3. Vessel obliteration
4. Pulmonary venous hypertension

World Health Symposium Classification

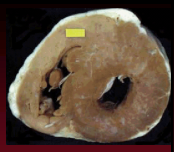
1. Pulmonary *arterial* (precapillary) HTN
2. Pulmonary venous HTN
3. Disorders of the respiratory system and/or hypoxemia (Lung Disease)
4. Chronic thrombotic or embolic disease
5. Unknown etiology/mechanism (Sarcoid, Sickle cell, metabolic disorders, Chronic hemolytic anemia)

Types of Pulmonary Hypertension



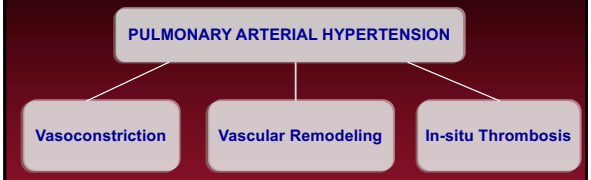
IPAH = idiopathic pulmonary artery hypertension; APAH = associated pulmonary artery hypertension.

Physiologic Consequences

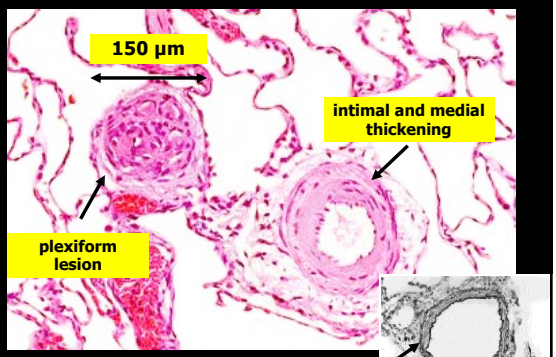


- Multiple causes lead to a *final common* pathway of disease
- Progressive distal arterial narrowing, medial hypertrophy and intimal thickening/fibrosis
- Loss of capacity to dilate and recruit unused pulmonary vasculature
- Impaired gas exchange
- Right ventricular hypertrophy and failure

PAH: Pathogenesis

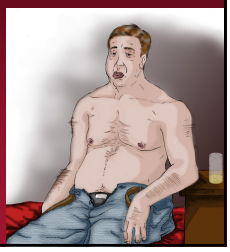


1. Humbert M et al. *N Engl J Med.* 2004;351:1425-1436.



Signs and Symptoms of PH

- Dyspnea, especially on exertion
- Fatigue
- Chest pain/discomfort
- Palpitations
- Pre-syncope/syncope
- Pitting Edema
- Ascites (Late stage)



Patient Evaluation: Goals

- Detect presence and severity of PAH
- Assess for underlying causes
- Guide therapy (Medical, Surgical, Cardiac)
- **Pre-capillary/capillary level**
- **Post-capillary causes** (Pulmonary venous and Cardiac disease)

Patient Evaluation: Imaging

- **Thoracic imaging** (Radiograph & CTA)
- Echo?
- V/Q scan?
- Cardiac MRI?
- **Right heart catheterization**
  - Assess PAP, PCWP and CO
  - Vasodilator trial (inhaled NO or intravenous flolan)
    - Positive response predicts:
      - Favorable response to Ca^{++} channel blockers
      - *Improved survival*

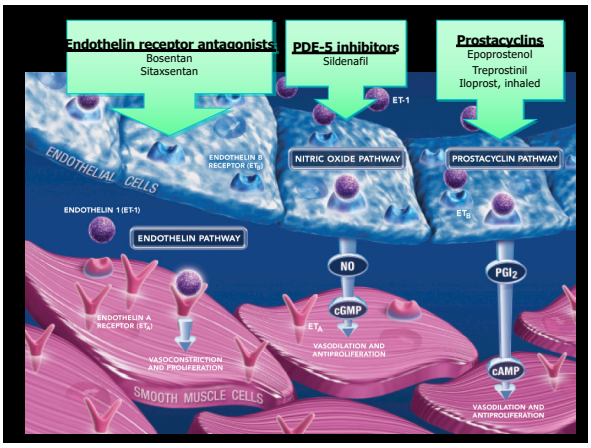
Current Therapeutic Considerations For ***Pre-Capillary*** Pulmonary HTN

Supplemental Oxygen
Diuretics
Anticoagulation
Calcium Channel Blockers

Prostacyclin derivatives
Endothelin-1 receptor blocking agents
Oral phosphodiesterase 5 (PDE5) inhibitors

Atrial Septostomy
Pulmonary Endarterectomy
Lung or Heart-Lung Transplantation

G. Allada, MD, OHSU



What You Can Offer The Clinician

For **dyspnea of unknown origin**, identify subtle findings of pulmonary hypertension

-Dilated/thickened right heart

-Interventricular septal straightening

- Bowing or Interatrial septum

-Enlarged pulmonary arterial diameter

-Reflux of contrast into the IVC

-Dilated Right Atrium (TR)

What You Can Offer The Clinician

For **known** Pulmonary Hypertension:

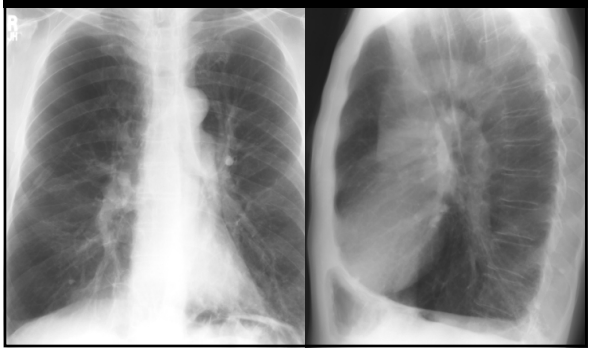
1. Assess for Left-sided heart disease
2. Chronic Thromboembolic Disease
3. Primary lung diseases – Chronic Hypoxia
4. Uncorrected congenital heart disease; ASD, VSD or PFO (MRI?)
5. PVOD and Pulmonary Capillary Hemangiomatosis

Radiographic Evaluation

- The size of pulmonary artery and right ventricle (Lateral view best).
- Presence of underlying lung disease.
- Presence of cardiac disease or chronic pulmonary venous hypertension.
- Associated findings: obese patient, TIPS shunt, prior cardiac surgery or shunt vascularity.

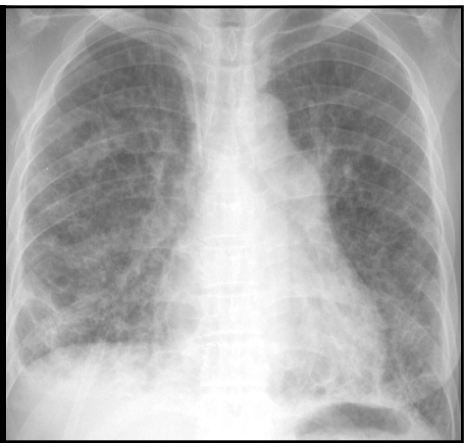
Emphysema and Pulmonary Hypertension

How is this different than Chronic Bronchitis?



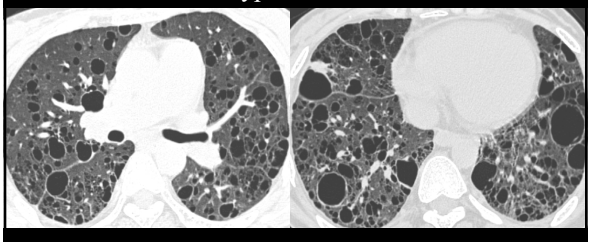
40 yo female with pulmonary hypertension receiving IV Prostacyclin therapy.

Why do you think she has Pulmonary hypertension?



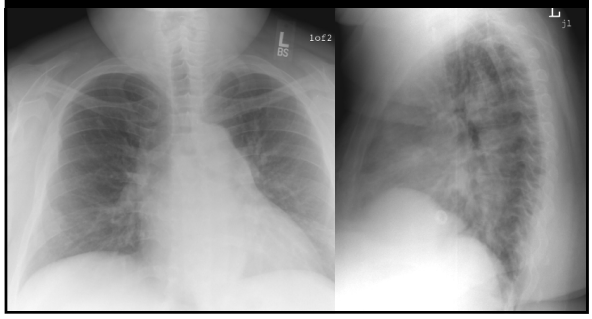
Lymphocytic Interstitial Pneumonitis from Autoimmune Disease

<u>Multifactorial</u>: Chronic Lung Disease/Hypoxia and potentially Autoimmune associated pulmonary hypertension

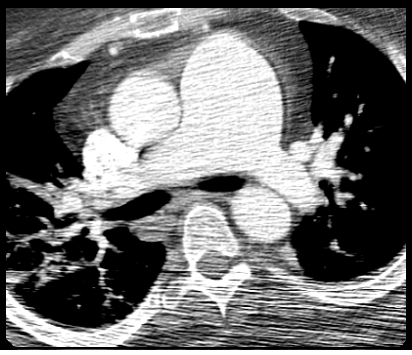


Enlarged Pulmonary Arteries

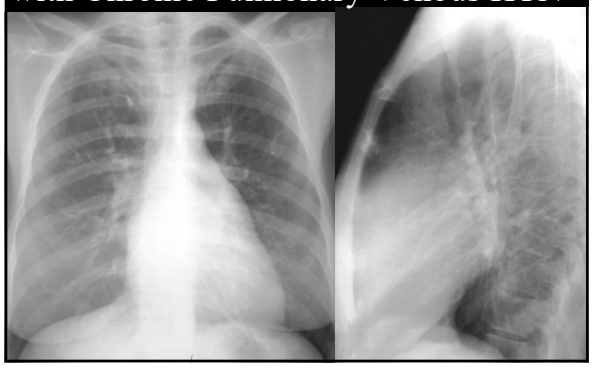
Probable Etiology?



Sleep Apnea/Chronic Hypoxia

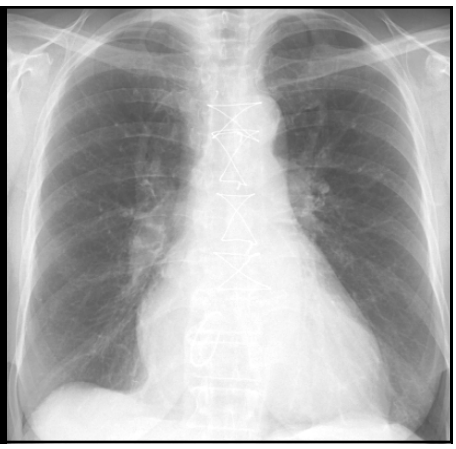


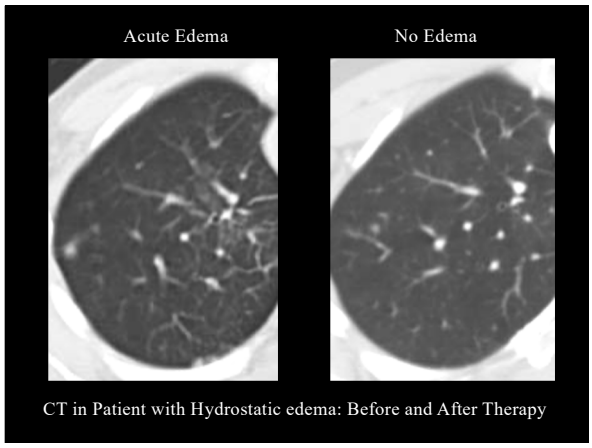
<u>Post Capillary Etiology</u>: Mitral Stenosis with Chronic Pulmonary Venous HTN



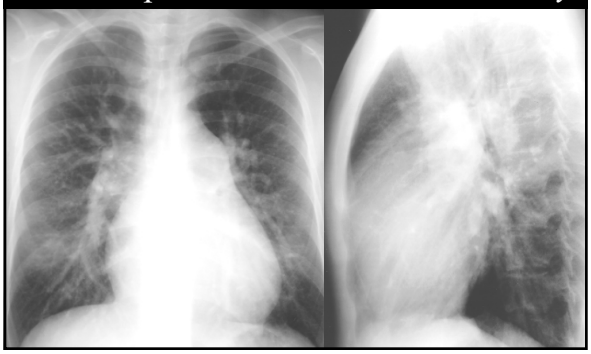
Mitral valve prosthesis for stenosis and tricuspid annuloplasty 5 years ago.

Why is this *vascular cephalization* still present?

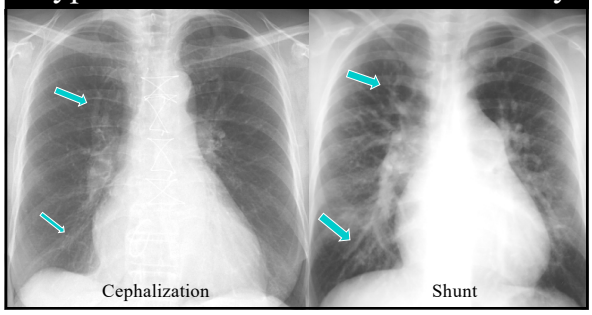




Pulmonary Hypertension Secondary to Atrial Septal Defect: *Shunt Vascularity*



Lower lobe vessels are helpful to distinguish between Chronic venous hypertension versus shunt vascularity



Number of nodding-off events per lecture (NOELs) per 100 attendees over length of time of presentation

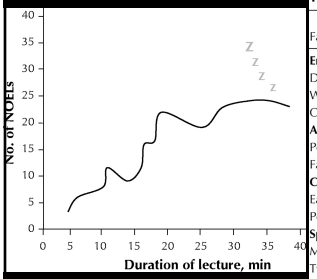


Table 1: Risk factors for nodding off at lectures

Factor	Odds ratio (and 95% CI
Environmental	
Dim lighting	1.6 (0.8–2.5
Warm room temperature	1.4 (0.9–1.6
Comfortable seating	1.0 (0.7–1.3
Audiovisual	
Poor slides	1.8 (1.3–2.0
Failure to speak into microphone	1.7 (1.3–2.1
Circadian	
Early morning	1.3 (0.9–1.8
Post prandial	1.7 (0.9–2.3
Speaker-related	
Monotonous tone	6.8 (5.4–8.0
Tweed jacket	2.1 (1.7–3.0
Losing place in lecture	2.0 (1.5–2.6

Note: CI = confidence interval

Rockwood, K. et al. CMAJ 2004;171:1443-1445

CMAJ • JAMC

Spiral CT Evaluation: Pulmonary Arterial Disease

- Supporting evidence of PAH: dilation of IVC, contrast reflux in IVC/hepatic veins and increased pulmonary arterial diameter
- Secondary cardiac changes: Right atrial and ventricular dilation/hypertrophy
- Bronchial arterial hypertrophy
- Mosaic lung attenuation

Normal Pulmonary CTA

- Contrast Dynamics:
- Transient interruption of contrast is common
- No *continuous* contrast reflux into IVC (<5cc/sec infusion rate)
- Anatomic:
- Interventricular septum convex to right
- Right ventricular anterior wall <4mm
- Pulmonary arterial diameter < 3 cm or same diameter as ascending aorta

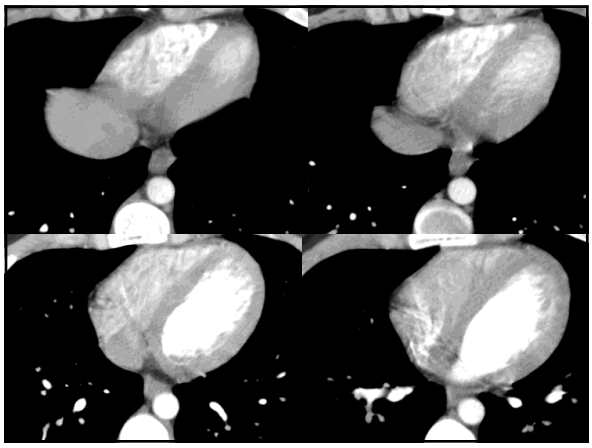
Contrast Dynamics: Transient Interruption of Contrast

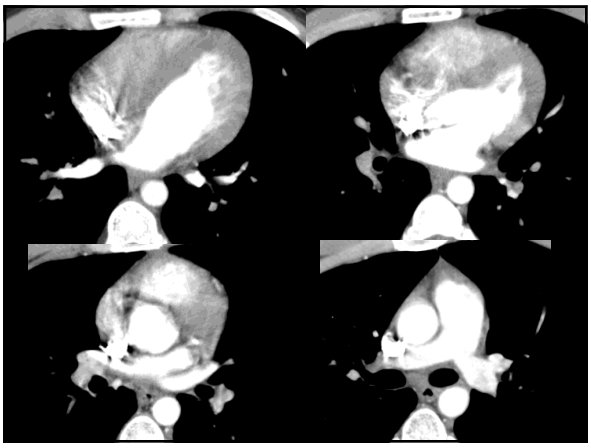
- Brief contrast interruption by unopacified blood entering the right atrium from the IVC *after deep inspiration*
- Often seen with *normal* cardiac output and right heart/pulmonary arterial pressures
- Indicative of good "*forward flow*" of blood
- **Variable** change in density

Transient Interruption of Contrast

- Interruption occurs at the *same* scan level with a uniform decrease in opacification of arteries *without expansion*
- **Assess right heart chambers on the immediate preceding images**
- Hyper-dense contrast will still be flowing in SVC (Contrast bolus did *not* finish)

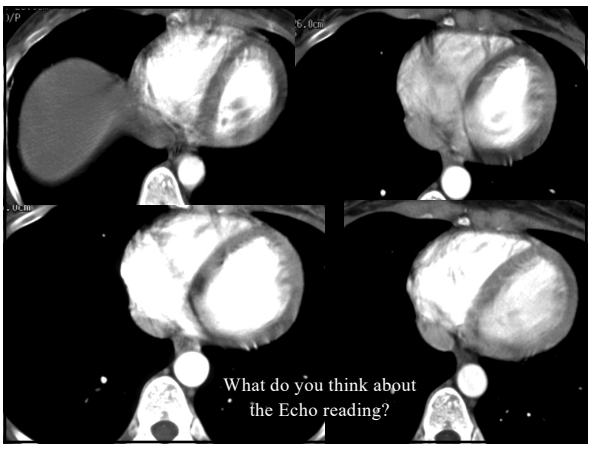
Gosselin MV, Rassner UA, Thieszen SL, Phillips J, Oki A. Contrast dynamics during CT pulmonary angiogram: analysis of an inspiration associated artifact. (2004) Journal of thoracic imaging. 19 (1): 1-7





Quick Case Example…

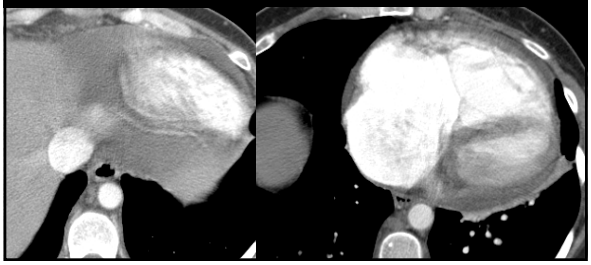
- ✓ 40 year old female referred for pulmonary arterial hypertension from an outside institution.
- ✓ Outside echocardiogram demonstrated systolic pressures of 60 mm Hg.



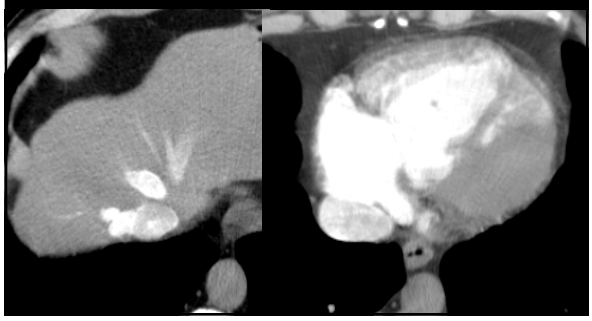
IVC Contrast Reflux

- ***Continuous* IVC** contrast reflux with venous dilation is associated with elevated right sided pressures, TR or reduced cardiac output (IV rate at 3 – 4cc/sec)
- *No IVC contribution* of unopacified blood into the right heart → ***Excellent pulmonary arterial opacification***

IVC Contrast Reflux With Right Ventricular Dilatation and Cardiac Rotation



Common Cardiac Changes



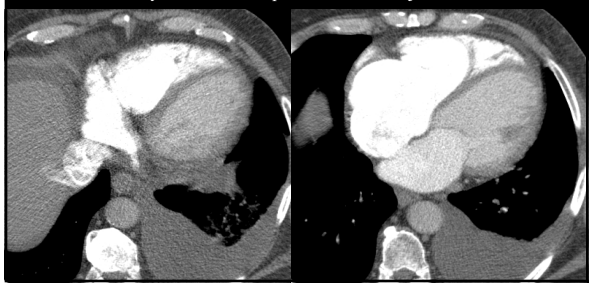
IVC Contrast Reflux

- Search for ancillary abnormalities
- **Constrictive pericardium:** Acute or chronic
- **Pulmonary artery:** Embolus and/or pulmonary hypertension
- **Cardiac:** Myocardial failure, septal defect or valvular abnormality

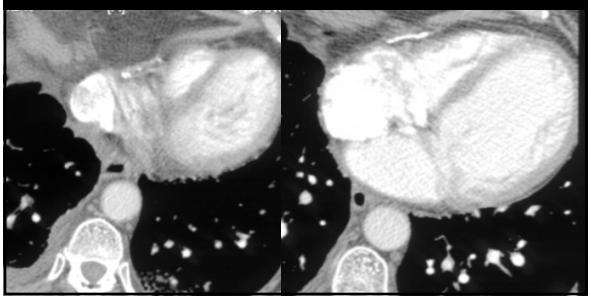
IVC Reflux From Dilated Cardiomyopathy: **EF 10-15 %**

Note the Poor opacification of LV and Aorta

Contrast Dynamics are helpful to estimate Ejection Fraction



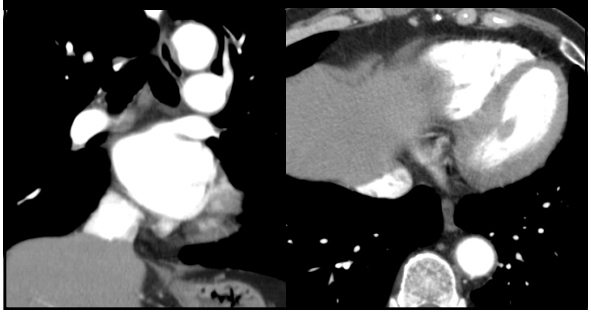
IVC Reflux: Constrictive Pericardium



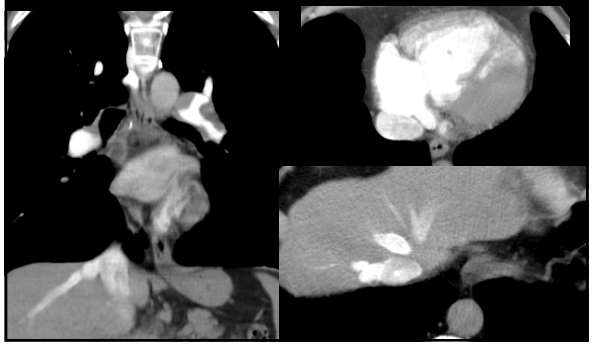
IVC Reflux and Pressure Correlation

- ***Mean*** pulmonary arterial pressures estimated by degree of reflux. (**Modified** by cardiac output)
- Extends above or below the diaphragm
- Below diaphragm correlates with ***mean*** pulmonary pressures **> 40mm Hg**
- Straightened or leftward bowing of Interventricular septal = ***systolic*** pressures **> 67 mmHg**

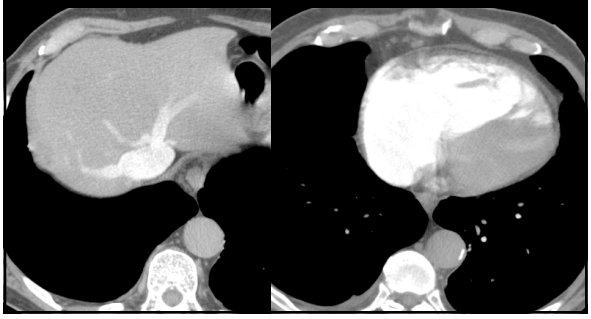
Mean Pulmonary Pressure: **35 mm Hg**



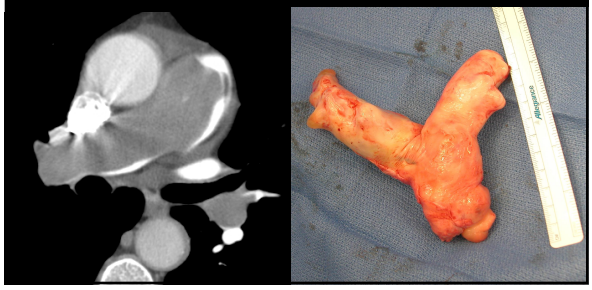
Mean Pulmonary Pressure: **70 mmHg**



IVC Reflux *With* Mild Right Ventricular Hypertrophy (Chronic)



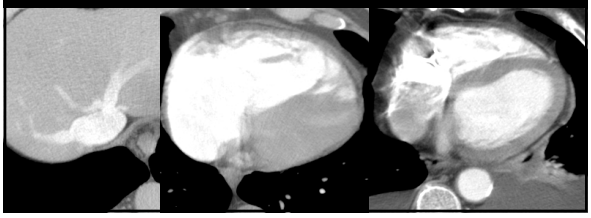
Pulmonary Arterial Sarcoma



Cardiac Changes and Contrast Reflux Are ***Dynamic***, Adjusting To Different Cardiopulmonary Conditions

CT Scan at Presentation

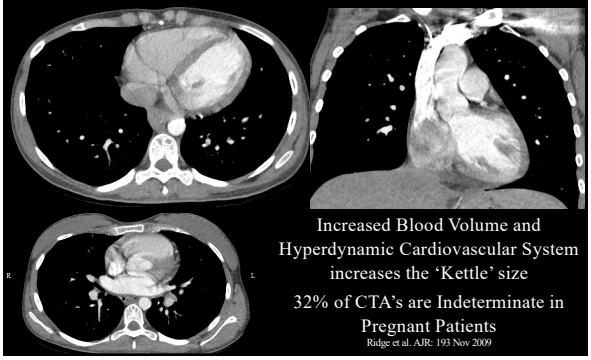
2 month F/up CT
Transient Interruption of Contrast



IVC Reflux and TIC Exceptions

- Cardiovascular capacity affects both IVC reflux of hyperdense contrast and the likelihood of TIC
- Large Cardiovascular capacity/Increased Blood Volume ***INCREASES TIC***
- Small Cardiovascular capacity/Petite persons ***INCREASES hyperdense contrast reflux***
- Like pouring the same amount of water into various sized Kettles: <u>One size doesn't fit all</u>!

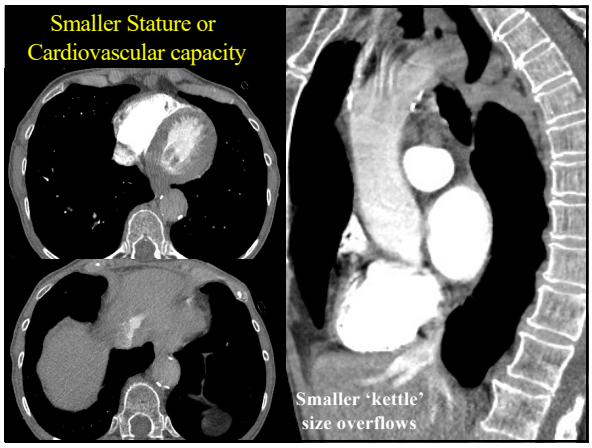
Pregnancy associated Transient Interruption of Contrast



Increased Blood Volume and Hyperdynamic Cardiovascular System increases the 'Kettle' size

32% of CTA's are Indeterminate in Pregnant Patients

Ridge et al. AJR: 193 Nov 2009



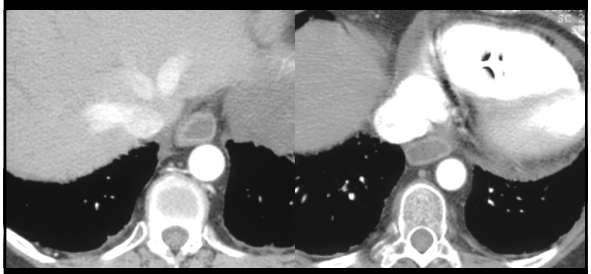
Spiral CT Evaluation: Etiology

- <u>**Primary pulmonary disease**</u>
  Parenchymal disease
  Airways disease
- <u>**Primary Cardiac disease**</u>
  Septal defects
  Myocardial failure
  Valvular disease
- <u>**Intrinsic Vascular Disease**</u>
  Thromboembolic Disease/Vasculitis/PPH

Note:

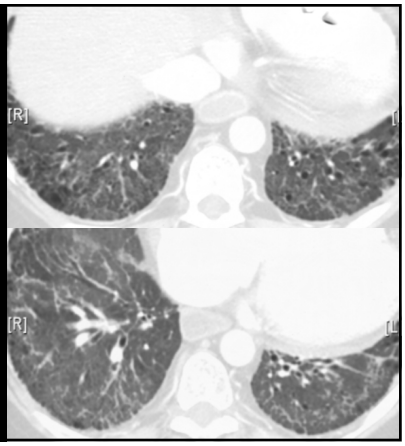
***More than one* of these causes may be present**

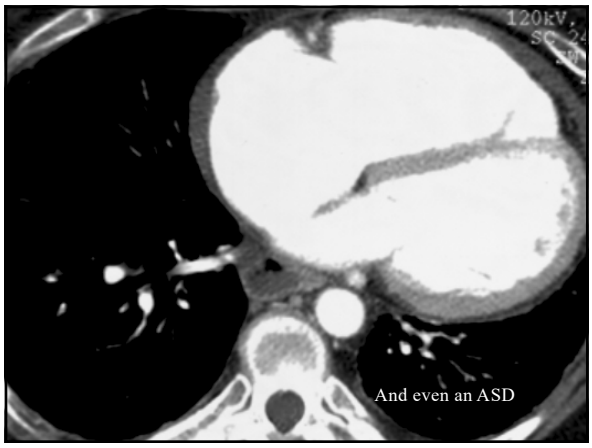
54 Year Old Women with Scleroderma



Scleroderma related pulmonary hypertension is very difficult to treat and can have multiple sources…

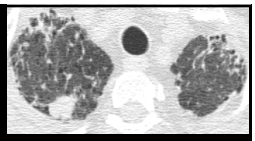
Chronic hypoxia from NSIP fibrosis *AND* intrinsic vascular disease are both commonly present



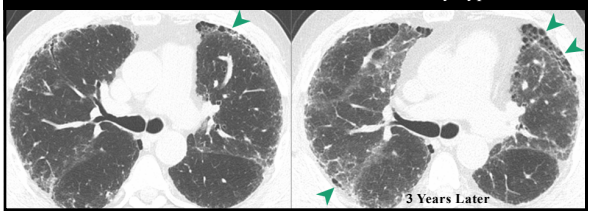


Usual Interstitial Pneumonitis (UIP):

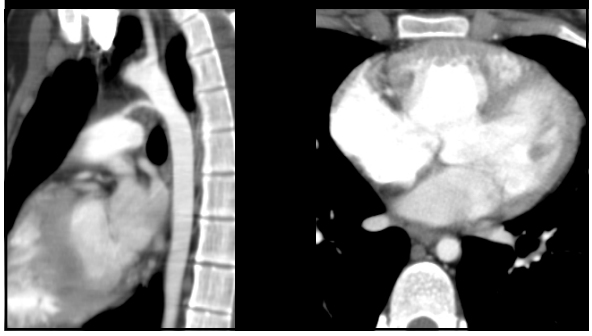
Three complications one should *ALWAYS* look for…

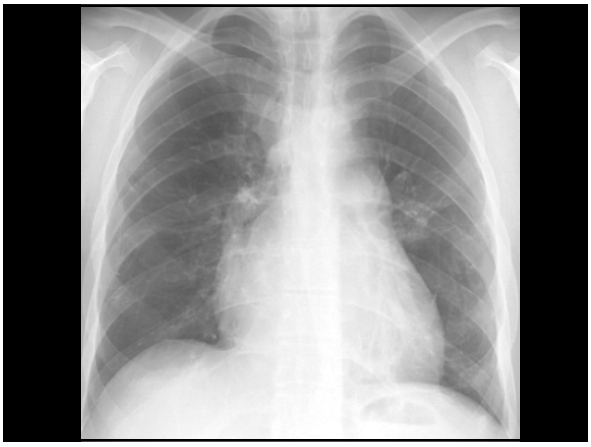


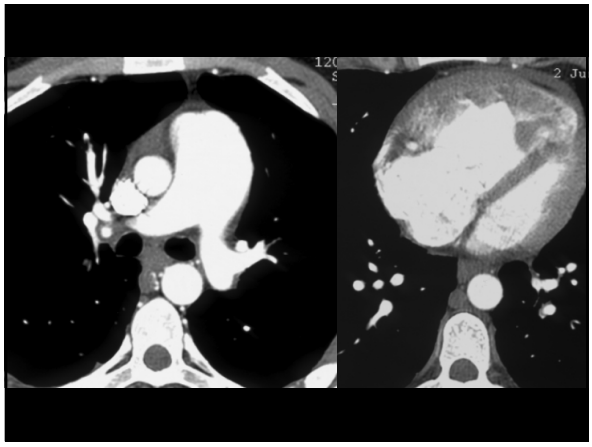
1. Any new nodule
2. New ground glass *without* evidence of fibrosis
3. Pulmonary hypertension



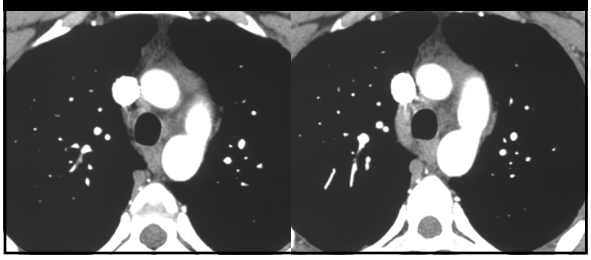
Patent Ductus Arteriosus and Ventricular Septal Defect



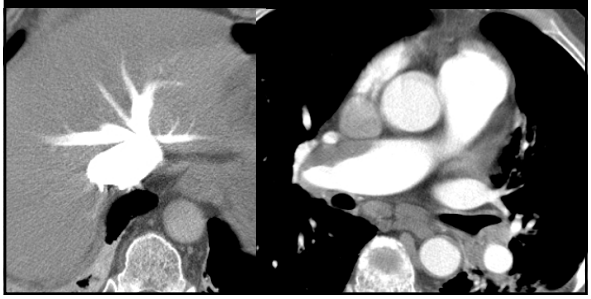




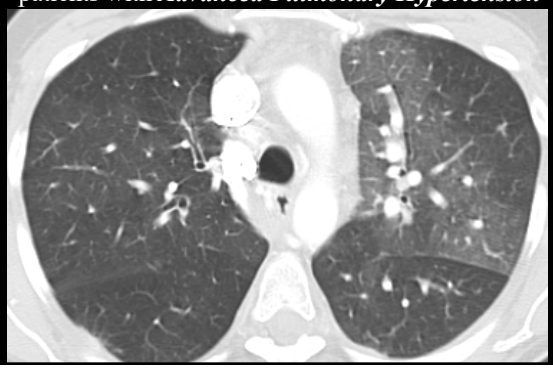
Diagnosis?

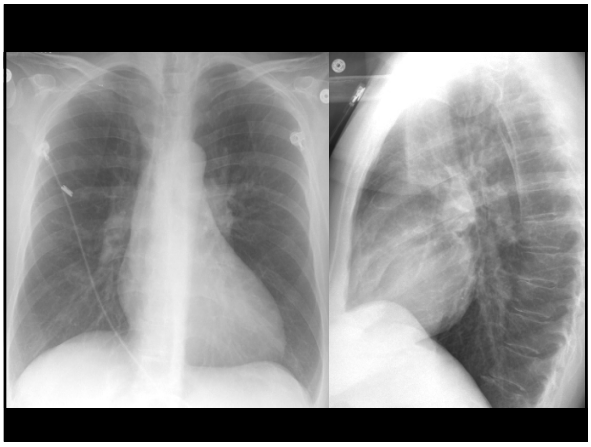


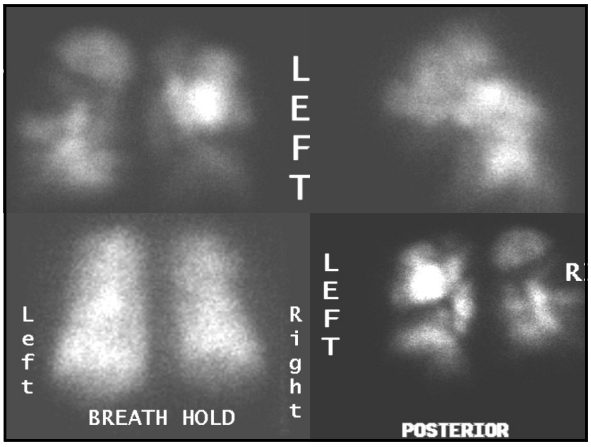
Chronic Thromboembolic Disease

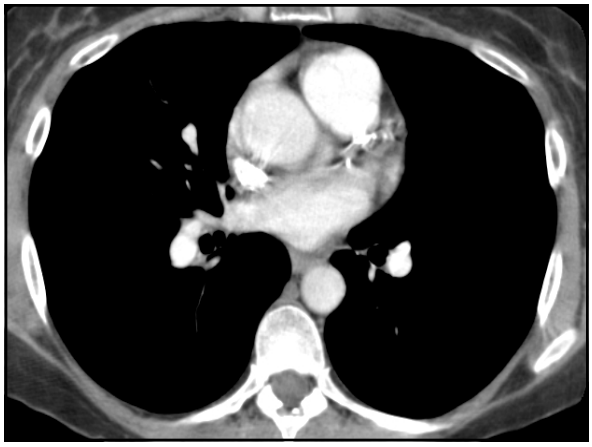


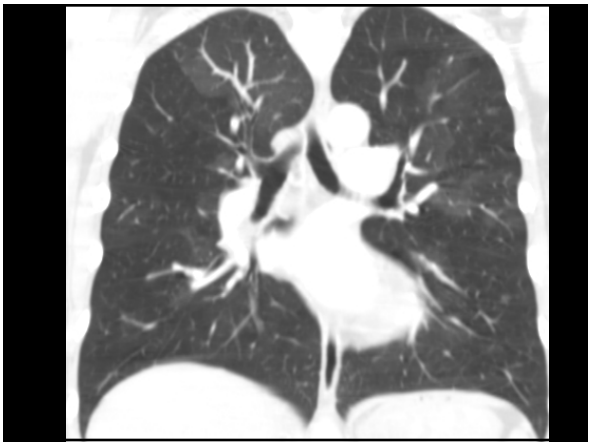
Mosaic Perfusion Pattern: Common in patients with ***Advanced Pulmonary Hypertension***

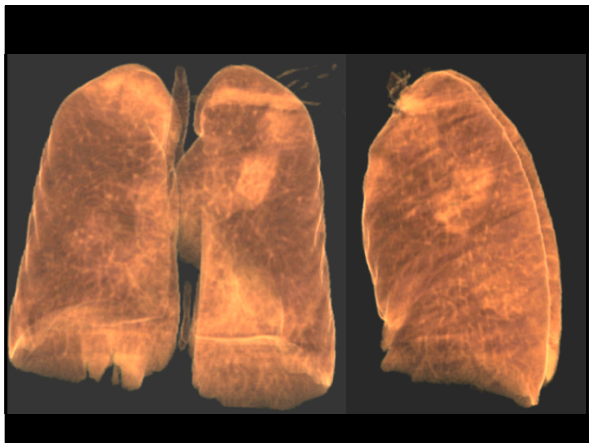




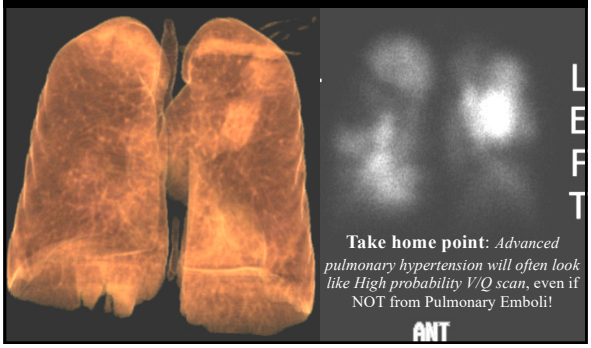






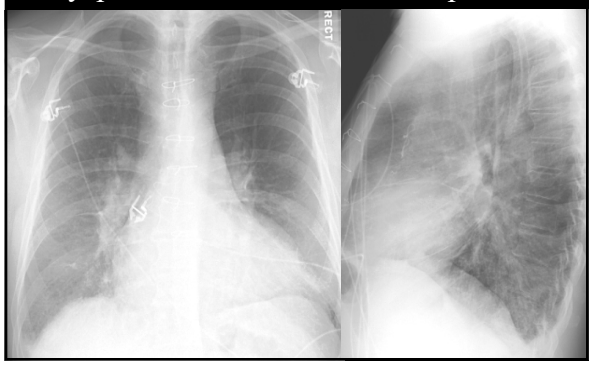


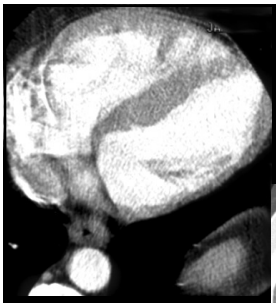
Mosaic Perfusion = Perfusion Defects



Take home point: *Advanced pulmonary hypertension will often look like High probability V/Q scan*, even if NOT from Pulmonary Emboli!

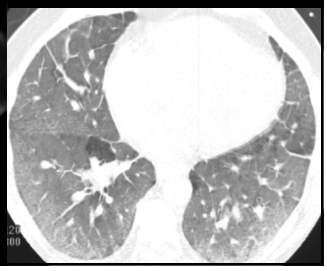
67 Year Old Male: Progressive Dyspnea and Recurrent CHF Episodes





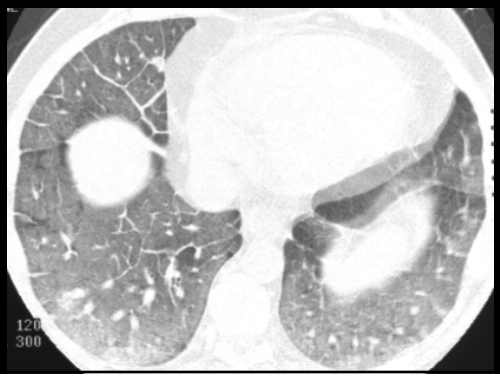
Moderate Pulmonary Hypertension with hypertrophy of RV wall.

Normal Left Ventricle Function and Pressures



Capillary Wedge Pressures were not increased…

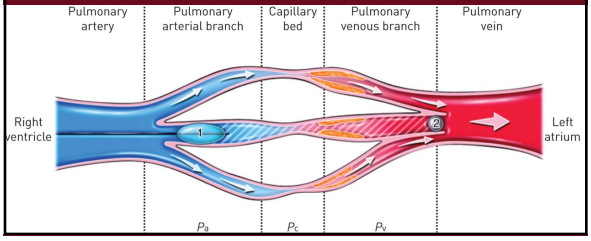
Probable Diagnosis? Report Impression?



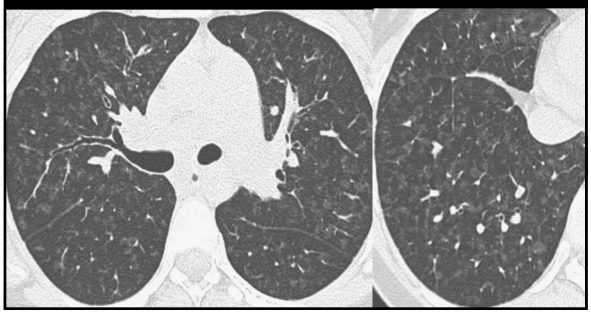
Why is the Wedge Pressure normal?

Pulmonary artery wedge pressures need a continuous column of blood and reflects the venous pressures in ***veins as or larger*** than where the balloon is located.

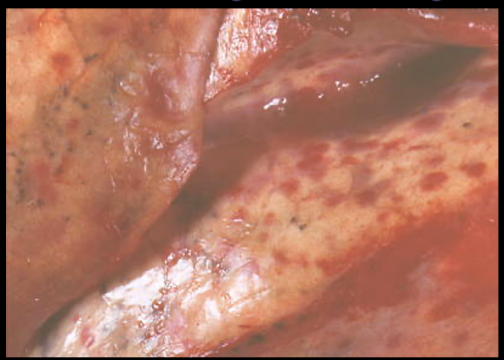
Pulmonary venoocclusive disease involves the very small veins with capillary proliferation, *but spares the larger veins*.



28 Year Old Female: Recent Diagnosis of Pulmonary Hypertension on Right heart Cath – Normal Left Ventricular Function



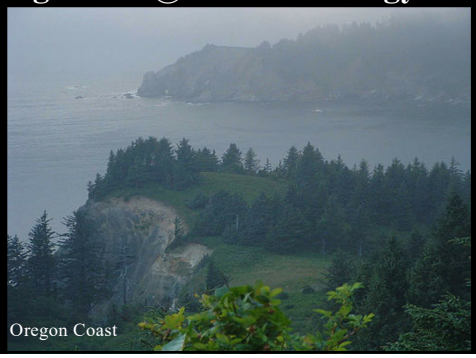
Pulmonary Capillary Hemangiomatosis: 5 – 10mm Hemorrhagic Nodules in Lung



Pulmonary Hypertension: **Summary**

- *Multiple etiologies* lead to a common final pathway of injury
- **Imaging Role**:
- Presence or absence of PAH
- **Etiology**: Most likely a primary pulmonary, primary cardiac, an intrinsic vascular/Thromboembolic disease…Or a combination?
- **Therapy is completely different for a *post capillary etiology!*

Thank you: gosselin@OHSU.edu or mgosselin@visionradiology.com



The imaging findings that is characteristic for transient interruption of contrast is?

- 1. Decreased opacification of the pulmonary arteries at the different levels
- 2. No contrast in the SVC with decreased pulmonary artery opacification
- 3. Reflux of hyperdense contrast into the IVC
- 4. Decreased opacification of pulmonary arteries with contrast in aorta and SVC

The imaging findings that is characteristic for transient interruption of contrast is?

Answer is #4: Decreased opacification of pulmonary arteries with contrast in aorta and SVC

Decreased pulmonary artery opacification with no contrast in the SVC means the bolus has run out, not necessarily TIC. Reflux of hyperdense contrast reflects poor cardiac forward flow or small cardiovascular capacity (Small heart and/or petite person). Decreased opacification at different levels reflects regional areas of decreased pulmonary flow and/or pulmonary emboli.

Gosselin MV, Rassner UA, Thieszen SL, Phillips J, Oki A. Contrast dynamics during CT pulmonary angiogram: analysis of an inspiration associated artifact. (2004) Journal of thoracic imaging. 19 (1): 1-7