

Chronic Thromboembolic Pulmonary Hypertension

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CHESTRAD 2023

A Case Review and Lecture Series
Saturday 15th July - Sunday 16th July - Monday 17th July
27 CPD Points

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Introduction

- Chronic thromboembolic disease is the only potentially curable cause of pulmonary hypertension (CTEPH)
- Characterized by organized thrombi causing pulmonary arterial narrowing or occlusion
- Leads to increased pulmonary vascular resistance

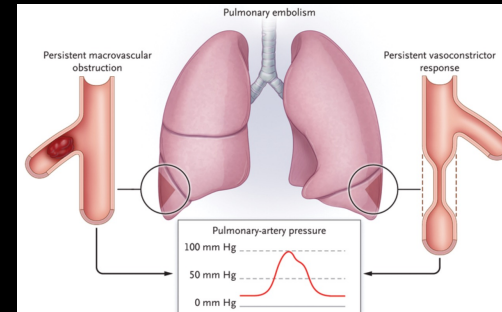
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Introduction

- Etiology of CTEPH is incompletely understood
- Roles of inflammation and vascular remodeling
- Risk factors
 - Splenectomy
 - Ventriculoatrial shunts in hydrocephalus
 - *Staphylococcus* infection
 - Chronic inflammatory disorders
 - Non-O blood groups

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Pathophysiology

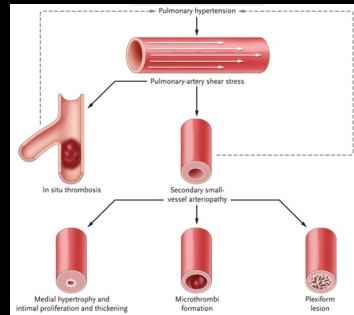


Piazza G. and Goldhaber SZ. *NEJM* 2011



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Pathophysiology

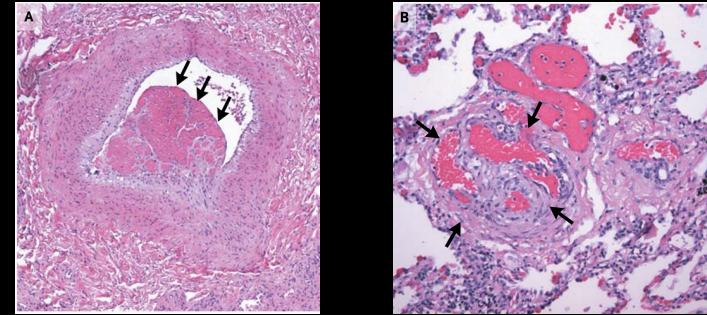


Piazza G. and Goldhaber SZ. *NEJM* 2011



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Histopathology



Piazza G. and Goldhaber SZ. *NEJM* 2011

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Objectives

- Define diagnostic evaluation for potential CTEPH
- Illustrate multimodality imaging features of CTEPH
- List some pitfalls that mimic CTEPH

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Diagnosis

- Clinical diagnosis may not be suspected because many patients do not report a history of acute PE.
- Imaging may begin with chest radiograph or echocardiogram
- CT often performed for abnormal TEE or radiograph

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CT Pulmonary Angiography

- Eccentric filling defects
- Webs
- Stenoses
- Occlusions



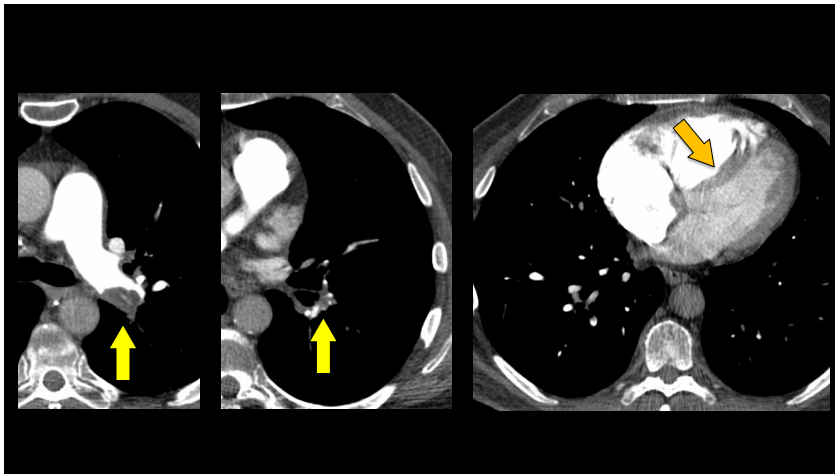
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CT Pulmonary Angiography

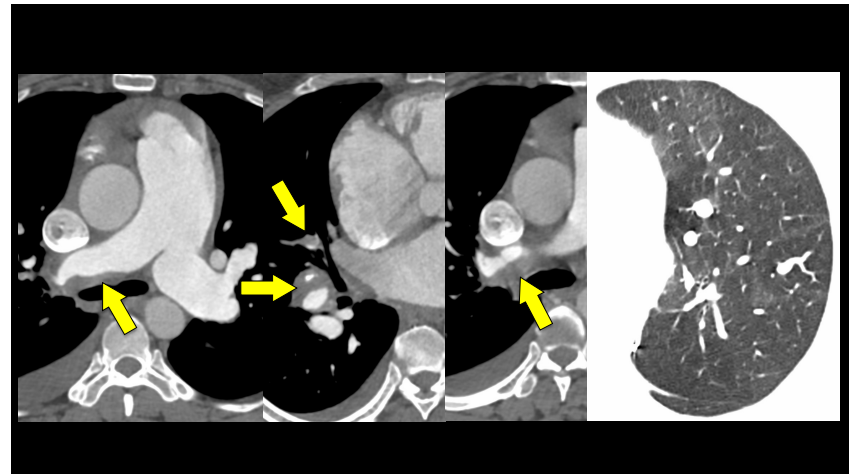
- RV enlargement and hypertrophy
- PA enlargement
- Bronchial artery hypertrophy
- Mosaic attenuation



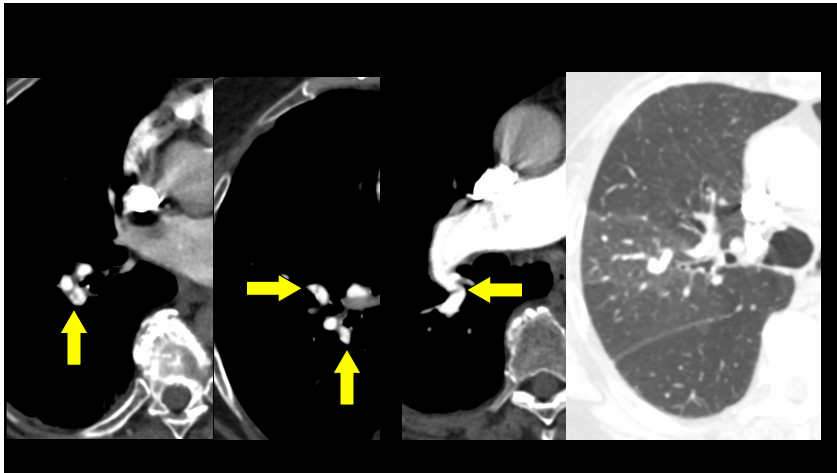
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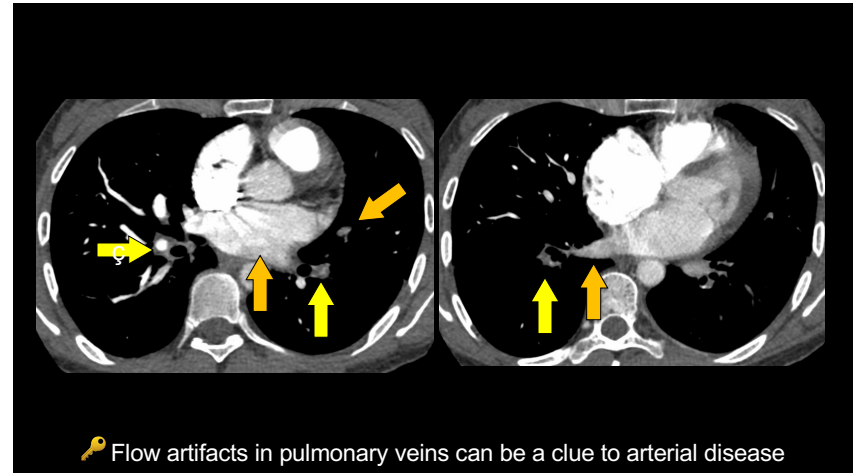
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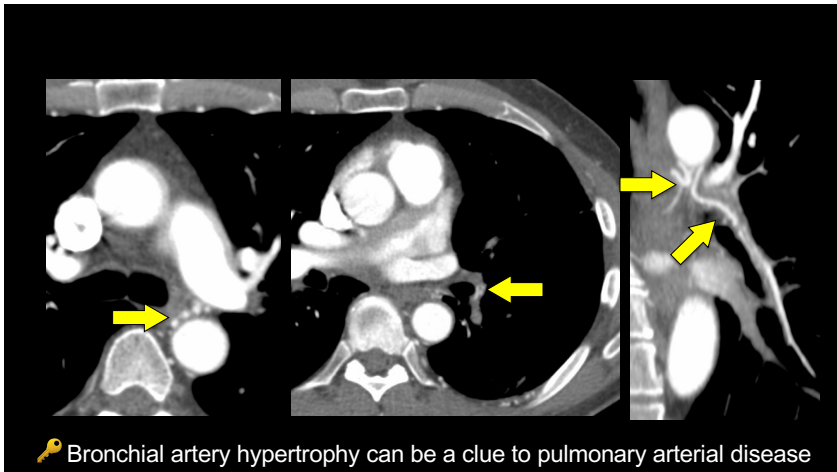


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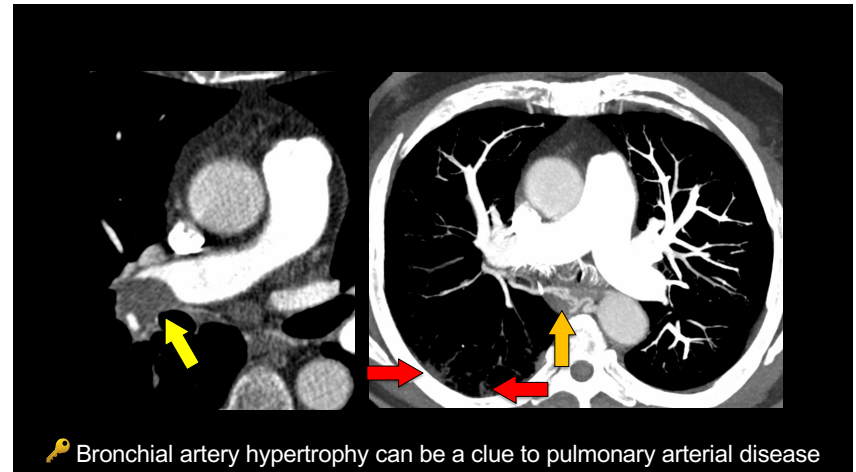
Flow artifacts in pulmonary veins can be a clue to arterial disease

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Bronchial artery hypertrophy can be a clue to pulmonary arterial disease

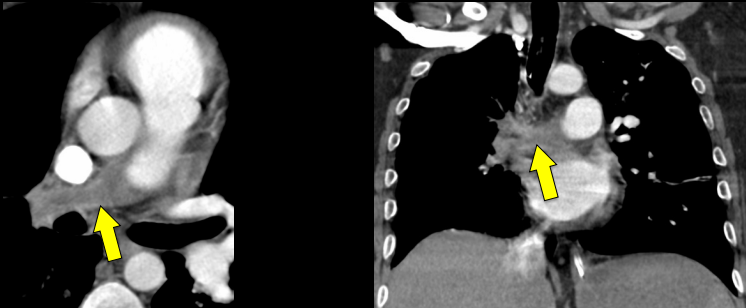
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Bronchial artery hypertrophy can be a clue to pulmonary arterial disease

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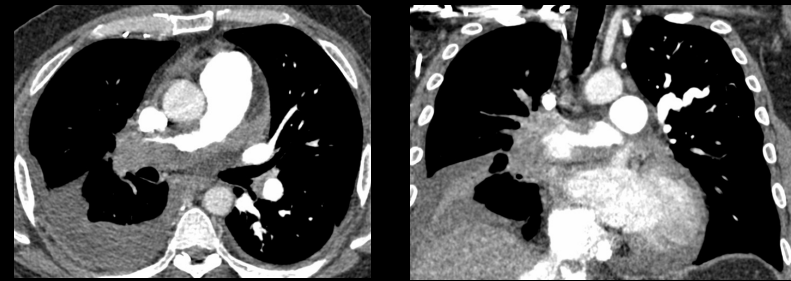
Atypical Presentation



Unilateral CTEPH is rare and should always raise the question of alternate diagnosis

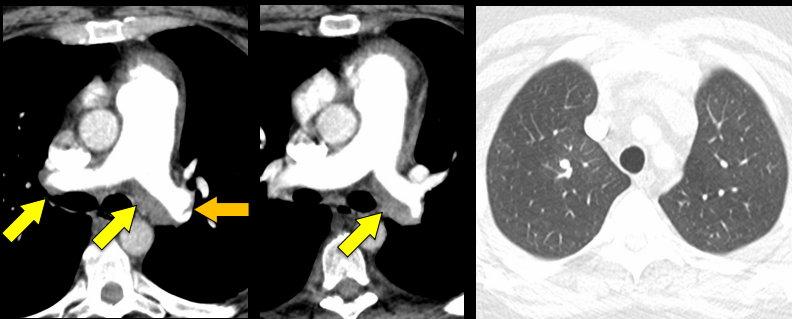
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Not a Mass

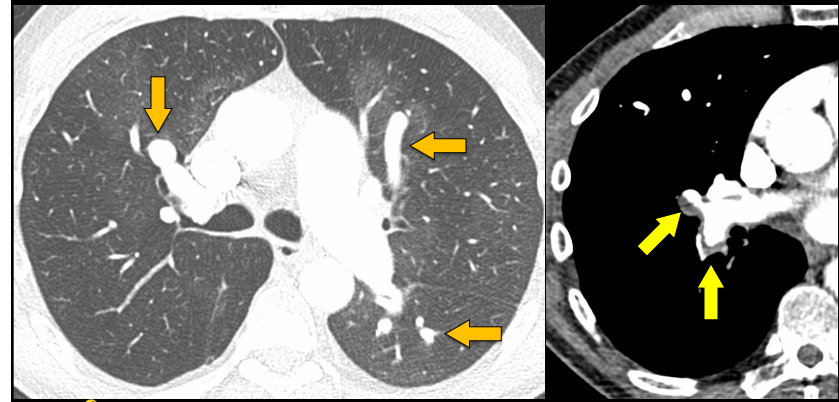


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Easy to Overlook



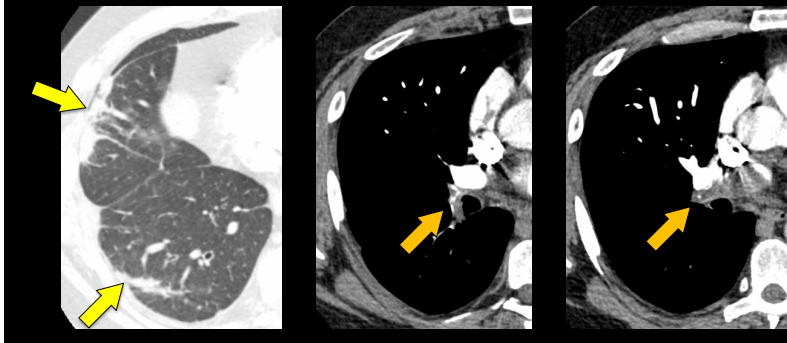
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Mosaic attenuation and vessel caliber heterogeneity need to be explained

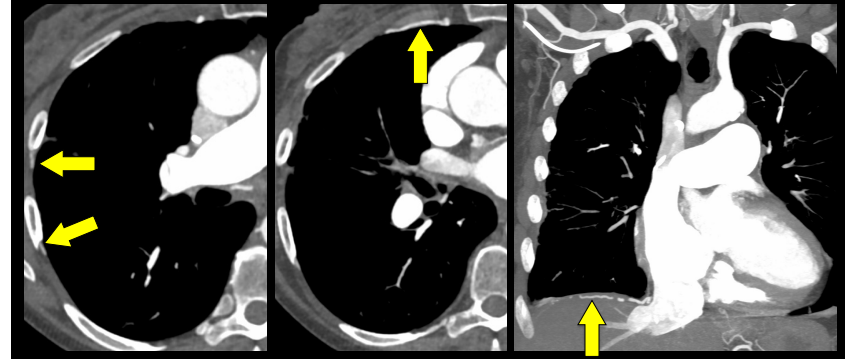
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Scar from Infarcts



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Transpleural Collaterals



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Transpleural Collaterals



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CT Pulmonary Angiography

- High accuracy for chronic PE when interpreted by subspecialty thoracic radiologist¹
- Lower diagnostic accuracy in many lower-volume settings
- Sensitivity 76% and specificity 96%²
- Sensitivity ↑ 88% at subsegmental level

¹Gopalan D et al. *Eur Respir Rev* 2017
²Dong C et al. *PLoS ONE* 2015

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Interobserver Agreement

Observer	1	2	3	4	5	6	Surgery
1	1	0.733	0.555	0.639	0.75	0.709	
2	0.714	1	0.611	0.484	0.627	0.682	0.533
3	0.733	0.611	1	0.515	0.586	0.729	0.53
4	0.555	0.484	0.515	1	0.536	0.525	0.384
5	0.639	0.627	0.586	0.536	1	0.609	0.502
6	0.75	0.682	0.729	0.525	0.609	1	0.541
Surgery	0.709	0.533	0.53	0.384	0.502	0.541	1

163 patients
Kligerman S. et al. 2023 (personal communication)

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Interobserver Agreement

Observer	1	2	3	4	5	6	Surgery
1	1	0.733	0.555	0.639	0.75	0.709	0.709
2	0.714	1	0.611	0.484	0.627	0.682	0.533
3	0.733	0.611	1	0.515	0.586	0.729	0.53
4	0.555	0.484	0.515	1	0.536	0.525	0.384
5	0.639	0.627	0.586	0.536	1	0.609	0.502
6	0.75	0.682	0.729	0.525	0.609	1	0.541
Surgery	0.709	0.533	0.53	0.384	0.502	0.541	1
+CT/year	500	10	100	50	20	200	

163 patients Kligerman S. et al. 2023 (personal communication)

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Interobserver Agreement

Observer	1	2	3	4	5	6
Slice thickness < 1 mm	0.602	0.338	0.361	0.418	0.355	0.362
Slice thickness > 1 mm	0.327	0.286	0.062	0.02	0.281	0.055

Segmental and subsegmental disease

Kligerman S. et al. 2023 (personal communication)

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V/Q Scanning

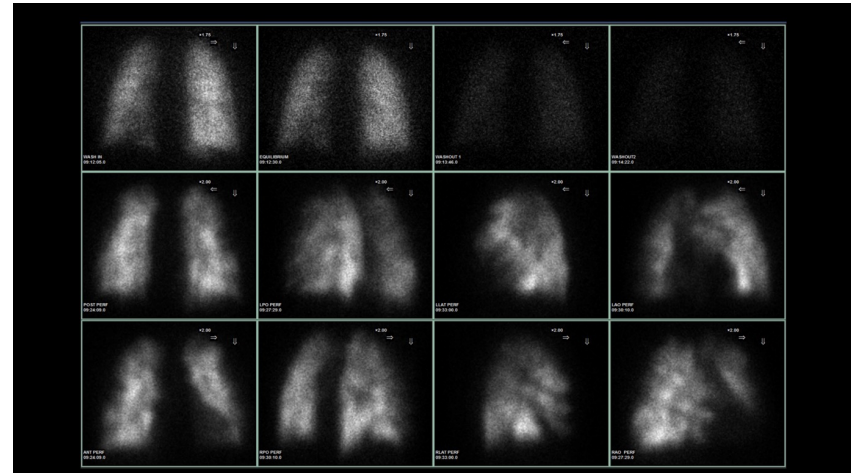
- V/Q scanning (especially with SPECT) is still considered reference standard
- More sensitive than CT for distal disease
- Many centers now use V/Q scanning and CT pulmonary angiography has complementary tests

Delcroix M et al. *Eur Resp J.* 2021

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MR Angiography

- MR angiography used in selected centers for acute pulmonary embolism evaluation
- Cardiac MRI used to evaluate right ventricular function in patients with pulmonary hypertension
- CMR and MRA can be used to evaluate patients after treatment

Kreitner K-FJ et al. *Radiology* 2004
 Kreitner K-FJ et al. *Eur Radiol* 2007
 Ley S et al. *Eur Radiol* 2012

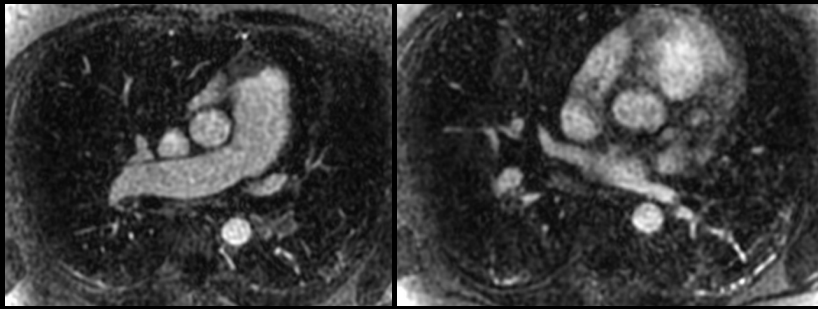
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MR Angiography

- | | |
|-------------------------|------------------------------|
| • Advantages | • Disadvantages |
| – Multiphase imaging | – Reduced spatial resolution |
| – Perfusion imaging | – Longer exam time |
| – Combine with CMR | – Access |
| – No ionizing radiation | – Lack of expertise |

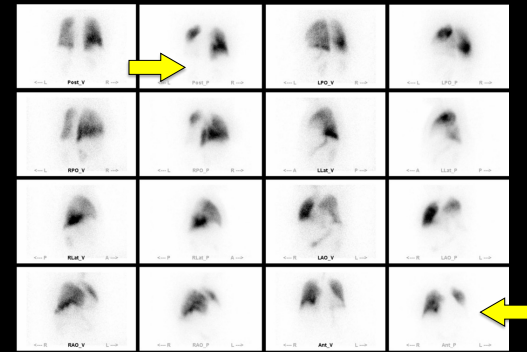
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MR Angiography



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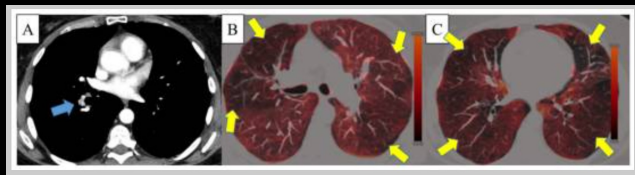
MR Angiography



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Dual Energy CT

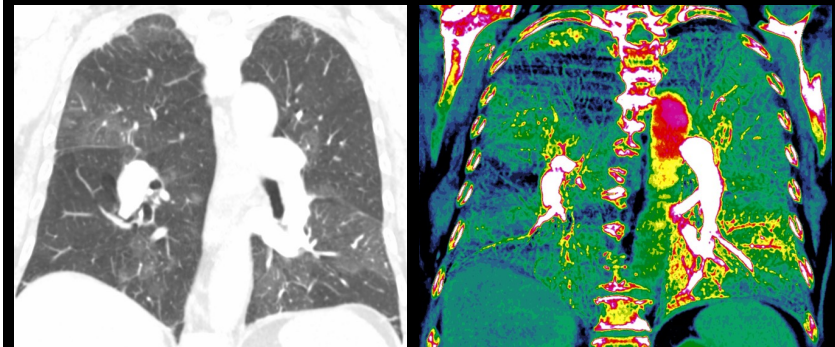
- Highly predictive of anatomic level of disease
- Moderate interobserver agreement
- Extent of perfusion defects not associated with level of disease or hemodynamic changes after surgery



Eberhard M et al. *Diagnostics (Basel)*. 2022

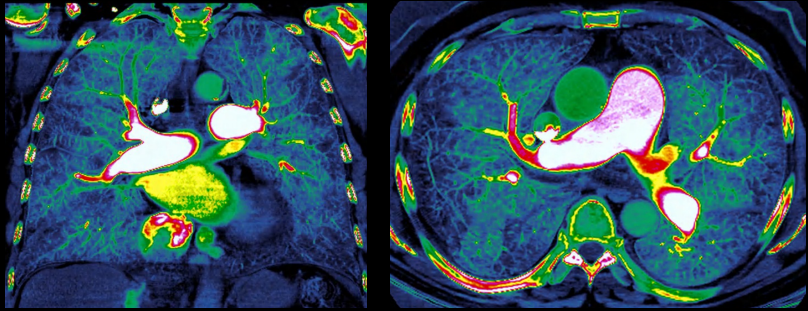
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Dual Energy CT



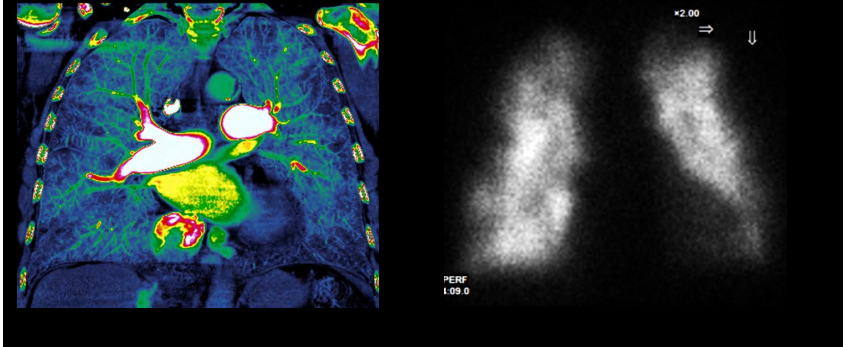
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Dual Energy CT



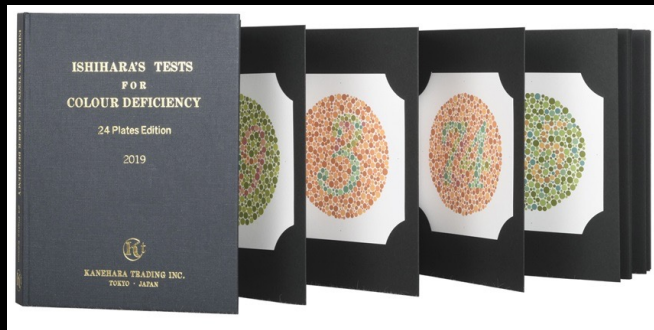
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Dual Energy CT



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Dual Energy CT



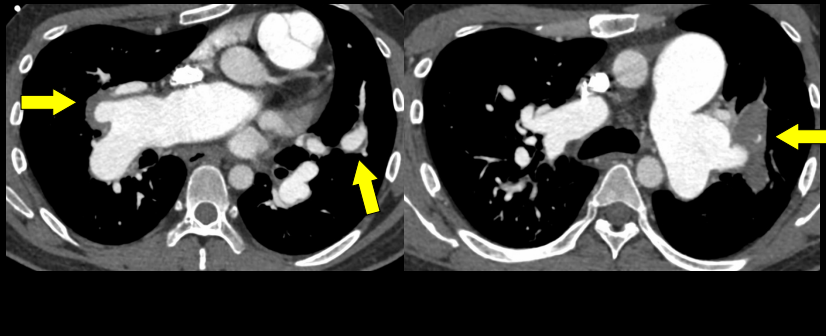
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Pitfalls



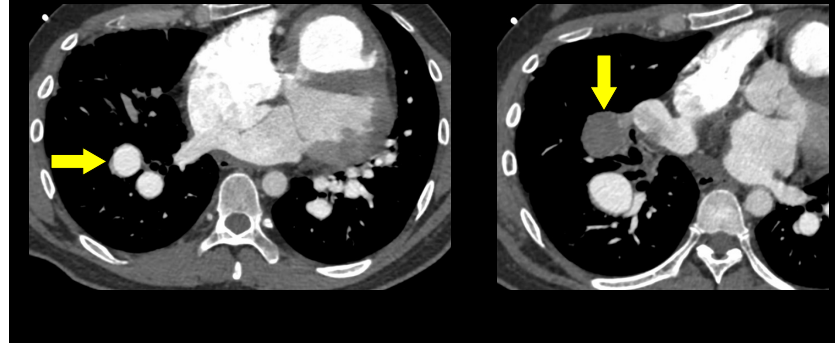
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In Situ Thrombus



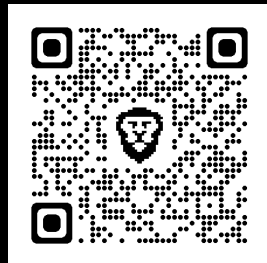
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In Situ Thrombus



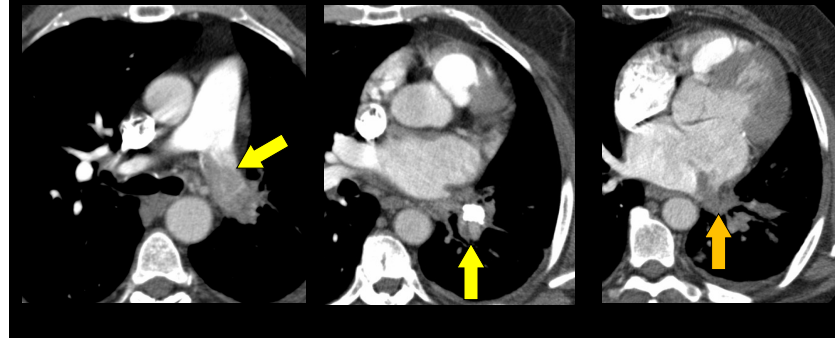
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Flow Artifacts



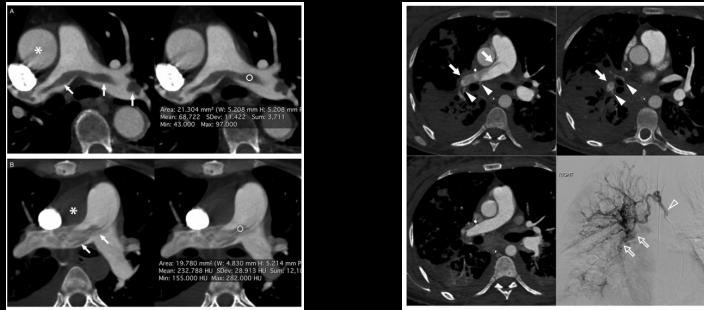
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Flow Artifacts



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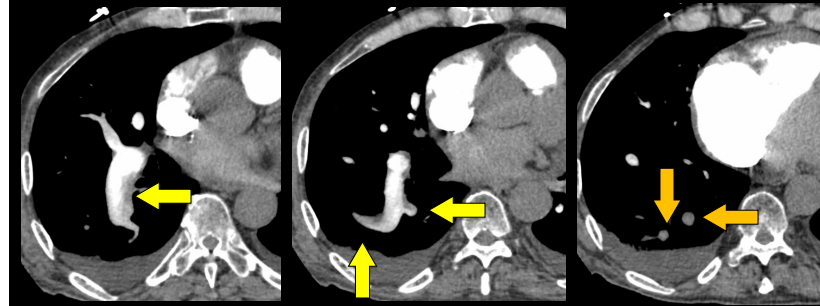
Flow Artifacts



Henry TS et al. *J Thorac Imaging* 2019

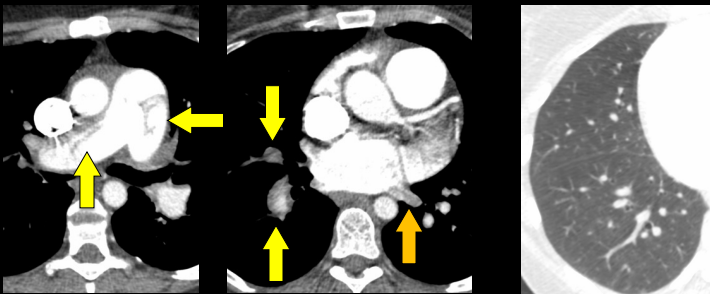
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Flow Artifacts



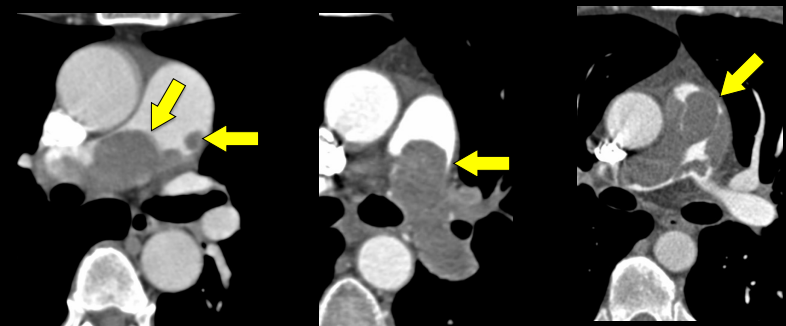
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Flow Artifacts



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Pulmonary Artery Sarcoma



🔑 Think sarcoma when you see the "world's largest PE"

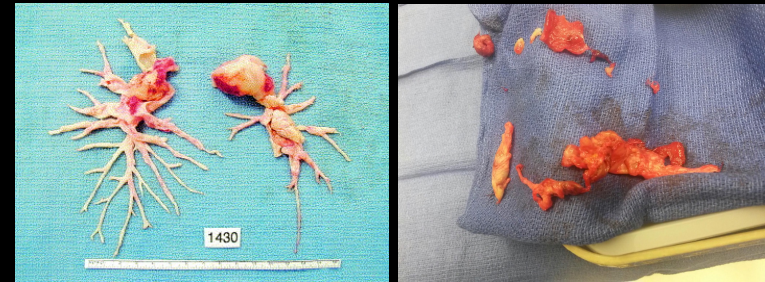
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Treatment

- CTEPH referral center
- Pulmonary endarterectomy
- Balloon pulmonary angioplasty
- PH management and lifelong anticoagulation

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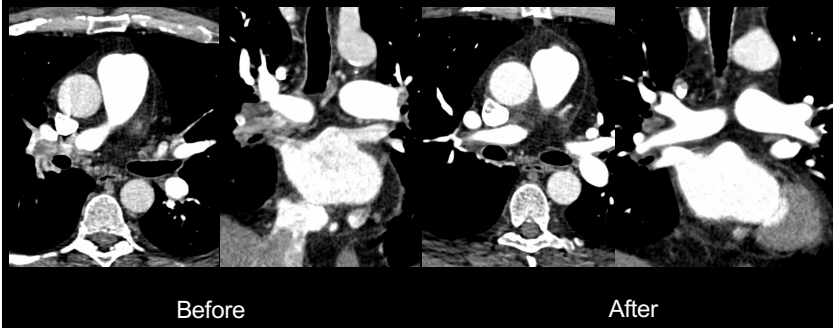
Endarterectomy



Jamieson SW et al. *Ann Thorac Surg* 2003

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Endarterectomy



Before

After

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Summary

- Chronic PE may be overlooked on routine imaging.
- Knowledge of the various manifestation of chronic PE can help the radiologist identify this potentially curable form of pulmonary hypertension.
- Awareness of pitfalls can help avoid misdiagnosis.

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