CTEPH EHK KROONILINE TROMBEMBOOLILINE PULMONAALHÜPERTENSIOON RADIOLOOGI PILGU LÄBI

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Radioloogia I aasta

HUVIDE KONFLIKTID

- Siemens Healthineers GmbH väärtpaberid alates 05.2022

MÕISTED

- Pulmonaalhüpertensioon/pulmonaalarteri hüpertensioon (PAH) – rahuolekus kopsuarteri keskmine rõhk (mPAP) üle 20 mmHg (ehk 21 mmHg ja enam)
  - Määramiseks sobivad nii mitteinvasiivsed (transtorakaalne ehhouuring) kui invasiivsed (RHC ehk parema südamepoole kateteriseerimine) meetodid
- CTEPD – krooniline trombembooliline kopsuhaigus, ERS seisukoha järgi sümptomaatilised patsiendid posttrombootiliste muutustega kopsuarterites koos või ilma PH-ta
- CTEPH – eelmine mõiste konkreetselt koos PH-ga
- DECT – kahe energiaga kompuutertomograafia
- DS-DECT – kahe allikaga (röntgentoruga) kahe energiaga kompuutertomograafia
- PBV – kopsu veremaht (*pulmonary perfused blood volume*), olemuselt kvalitatiivne mõiste

NICE KLASSIFIKATSIOON 2013

1. PAH
   - 1.1 Idiopathic PAH
   - 1.2 Heritable PAH
     - 1.2.1 *BMPR2*
     - 1.2.2 *ALK1*, *ENG*, *SMAD9*, *CAV1*, *KCNK3*
     - 1.2.3 Unknown
   - 1.3 Drug and toxin induced
   - 1.4 Associated with:
     - 1.4.1 Connective tissue disease
     - 1.4.2 HIV infection
     - 1.4.3 Portal hypertension
     - 1.4.4 Congenital heart disease
     - 1.4.5 Schistosomiasis

1' PVOD and/or PCH

1" PPHN

2. PH due to left heart disease
   - 2.1 Left ventricular systolic dysfunction
   - 2.2 Left ventricular diastolic dysfunction
   - 2.3 Valvular disease
   - 2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

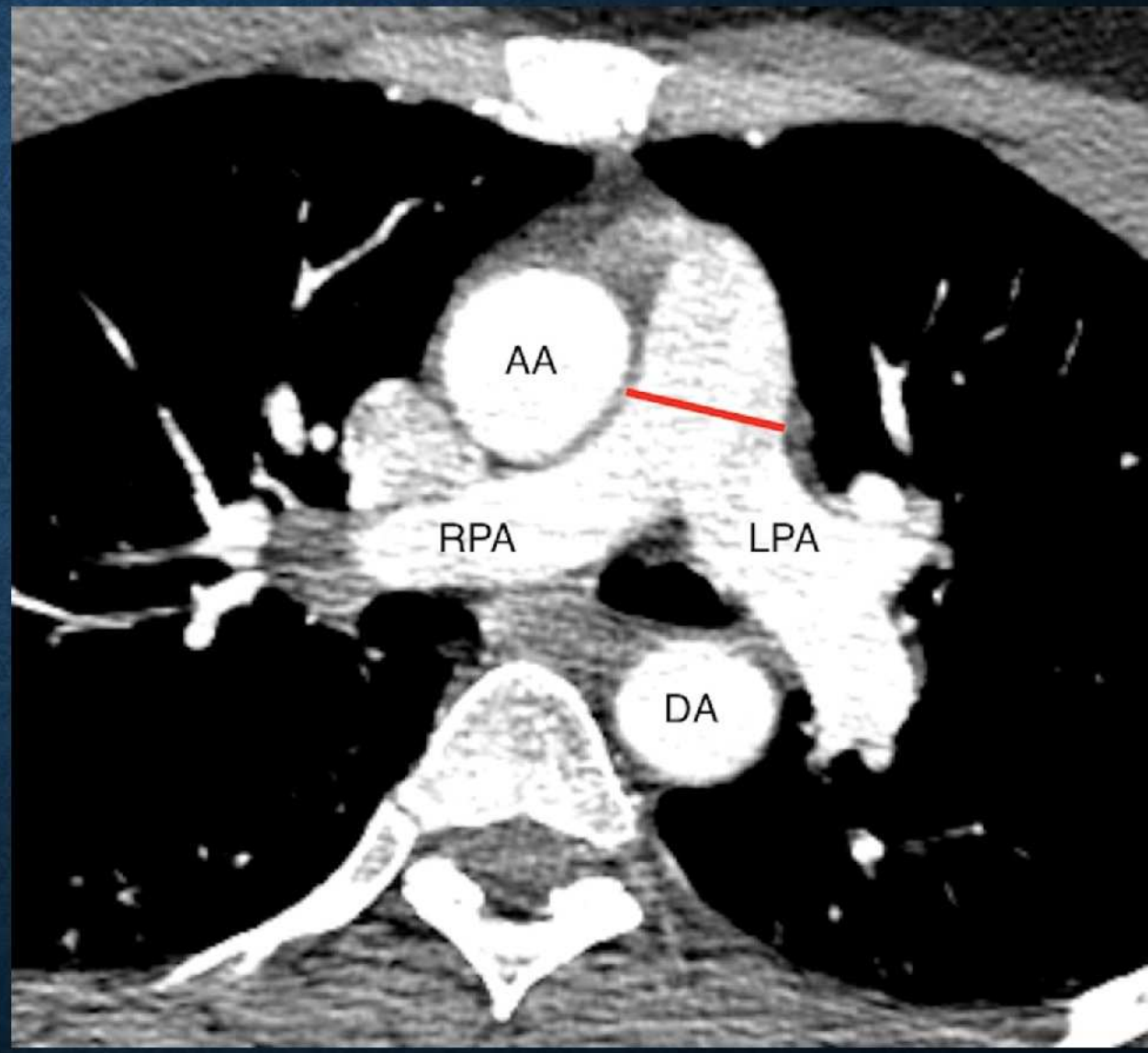
3. PH due to lung diseases and/or hypoxia
   - 3.1 COPD
   - 3.2 Interstitial lung disease
   - 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
   - 3.4 Sleep-disordered breathing
   - 3.5 Alveolar hypoventilation disorders
   - 3.6 Chronic exposure to high altitude
   - 3.7 Developmental lung disease

4. CTEPH

5. PH with unclear multifactorial mechanisms
   - 5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy
   - 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
   - 5.3 Metabolic disorders: glycogen-storage disease, Gaucher disease, thyroid disorders
   - 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

HAIGUSE DÜNAAMIKA

- Äge KATE, enamasti väljaravitav kaugtüsistusteta
- Osal juhtudest jäävad kroonilised muutused kopsuarteritesse
- Ägeda KATE järel CTEPH kujuneb välja 1 kuni 5% haigetest
- USA andmetel diagnoositakse alla 30% CTEPH juhtudest
- PAH-i vormidest ainuke (tinglikult) väljaravitav



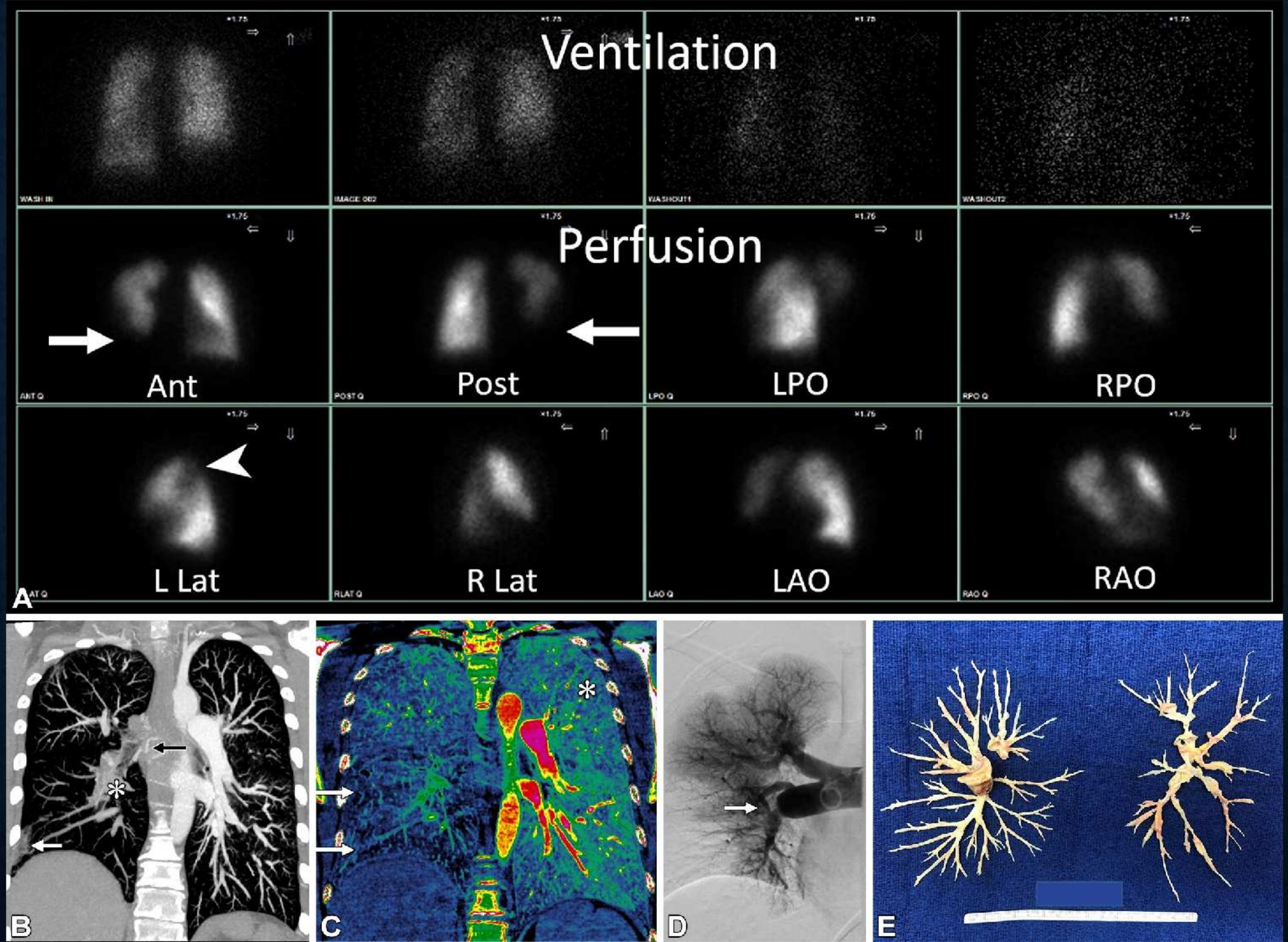
DIAGNOSTIKA

- Südame ultraheli (ehhouuring) – RV ülekoormuse hindamine, kaudne sPAP arvutamine, kambrite ja klappide anatoomia ja funktsioon
- Perfusioonuuring (V/Q skänn) – saab hinnata perfusioonidefekte (*mismatch*), tundlikkus ~97%, spetsiifilisus ~90-95%
  - Kirjanduse järgi valikuuring, kui 3 kuu pärast ravidoosis antikoagulatsiooniga inimene sümptomaatiline (düspnoe, koormustaluvuse langus jne)
  - Probleemiks mitteoklusiivse haiguse alahindamine
  - SPECT V/Q parem kui planaarne
- Kopsuarteri KT-angiograafia (sh DE-CTPA ehk kahe energiaga uuring)
- Parema südamepoole kateteriseerimine – invasiivne, võimaldab otseselt mõõta kopsuarteri rõhku ja eristada pre- ja postkapillaarset PAH-i, saab üle minna terapeutiliseks protseduuriks
  - DSPA – digitaalse subtraktsiooniga kopsuarteri angiograafia
- MRT, MRA ja MRT-perfusioon?

TÜÜPILINE DIAGNOSTILINE RADA

- Ehhouuring *kiirgusvaba*
- V/Q skänn *kiirgus*
- Kopsuarterite KT-angiograafia *kiirgus*
- RHC koos DSPA-ga *invasiivne ja kiirgus*
- Muud olulised lisauuringud (nt koronarograafia)

- Kokku vähemalt 4 uuringut ravi planeerimiseks!


Ventilation
Perfusion
Ant
Post
LPO
RPO
L Lat
R Lat
LAO
RAO
A
B
C
D
E

CTEPH Diagnosis*

Continue lifelong anticoagulation

Case Reviewed at CTEPH Multidisciplinary Conference

Cardiothoracic Surgeon specializing in Pulmonary Thromboendarterectomy (PTE)
Pulmonologist/Cardiologist specializing in pulmonary vascular disease
Cardiothoracic Radiologist
Interventionalist specializing in Balloon Pulmonary Angioplasty (BPA)

*outside referral

CTEPH excluded based on imaging and clinical data

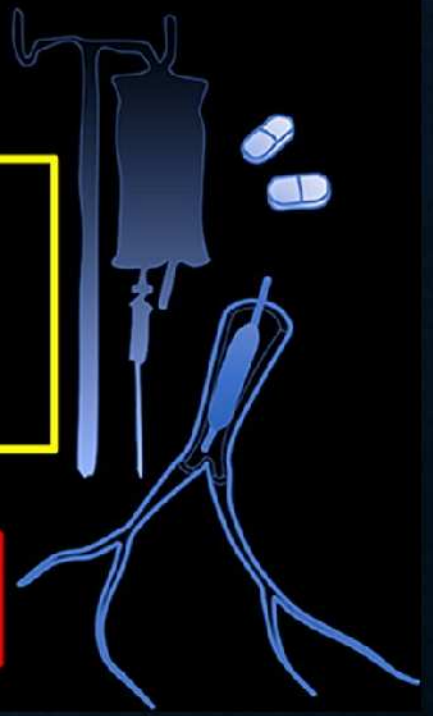
Treat the specific non-CTEPH condition

Technically operable

PTE

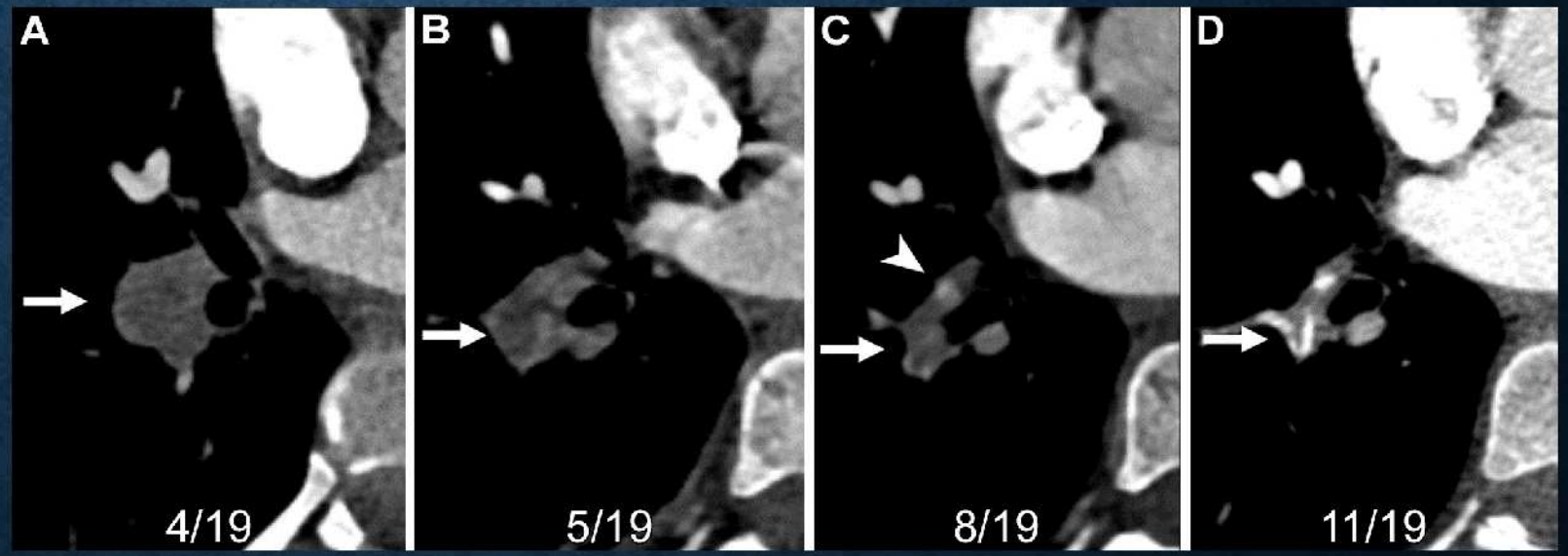
- Technically inoperable
- Technically operable but not operated (e.g. refusal, excess risk, risk to benefit)
- Residual PH s/p PTE

Targeted PH medical therapy
BPA consideration



KOPSUARTERITE KT-ANGIOGRAAFIA

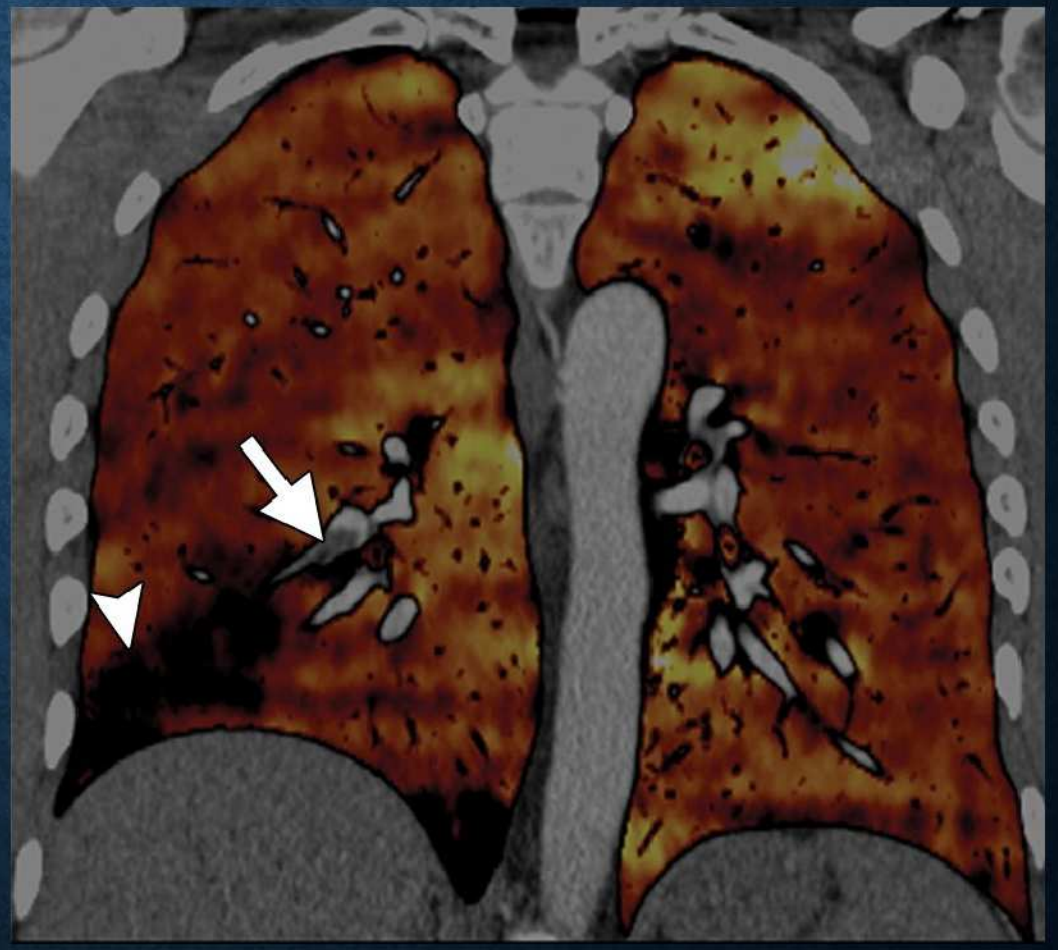
- Ühe energiaga(~kindel kVp, mAs modulatsioon) saame hinnata:
  - Kopsuarterite anatoomiat sh täitedefekte
  - Kopsukudet – alveolaartasandi ja interstitsiaalsed muutused
  - Pleurat ja rindkereseina
  - Südant (vähemalt parema poole)
  - Muud mis uuringuväljale jääb ja meile oluline

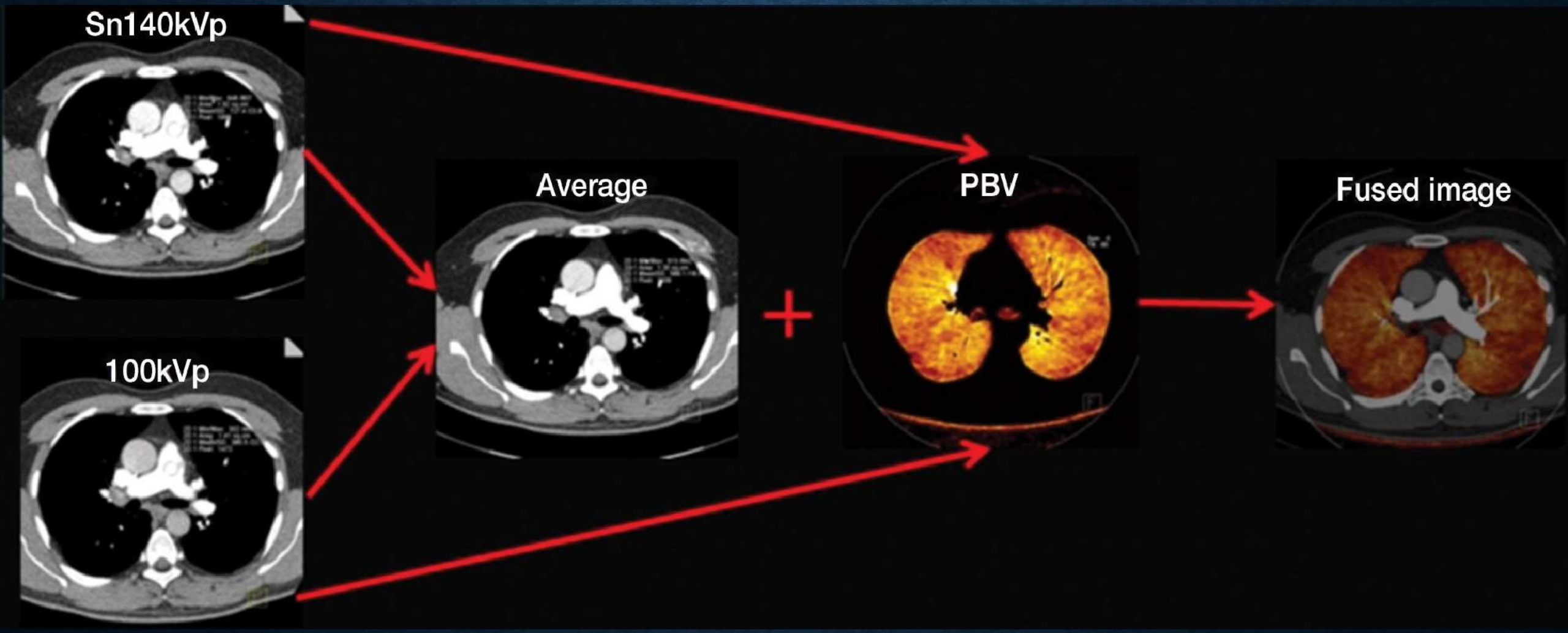


KAHE ENERGIAGA KT-UURING

Eelnevale lisaks saab:

- Hinnata regionaalset veremahtu (olemuselt *mitte tõeline* perfusioonuuring)
- Parandada subsegmentaarsete kopsuarterite nähtavust
- Vähendada lubjastustest tingitud (*beam hardening* ja *streak*-) artefakte
- Olemuselt DE-CTPA-st saab nii V/Q skänni kui tavalise CTPA kliinilist infot **ühe uuringuga hoides kokku nii raha kui aega**
- PBV korrelatsioon SPECT V/Q-ga varieerub
  - Tundlikkuse osas 83-96%
  - Spetsiifilisuse osas 76-99% (erinevus artefaktide tõttu)



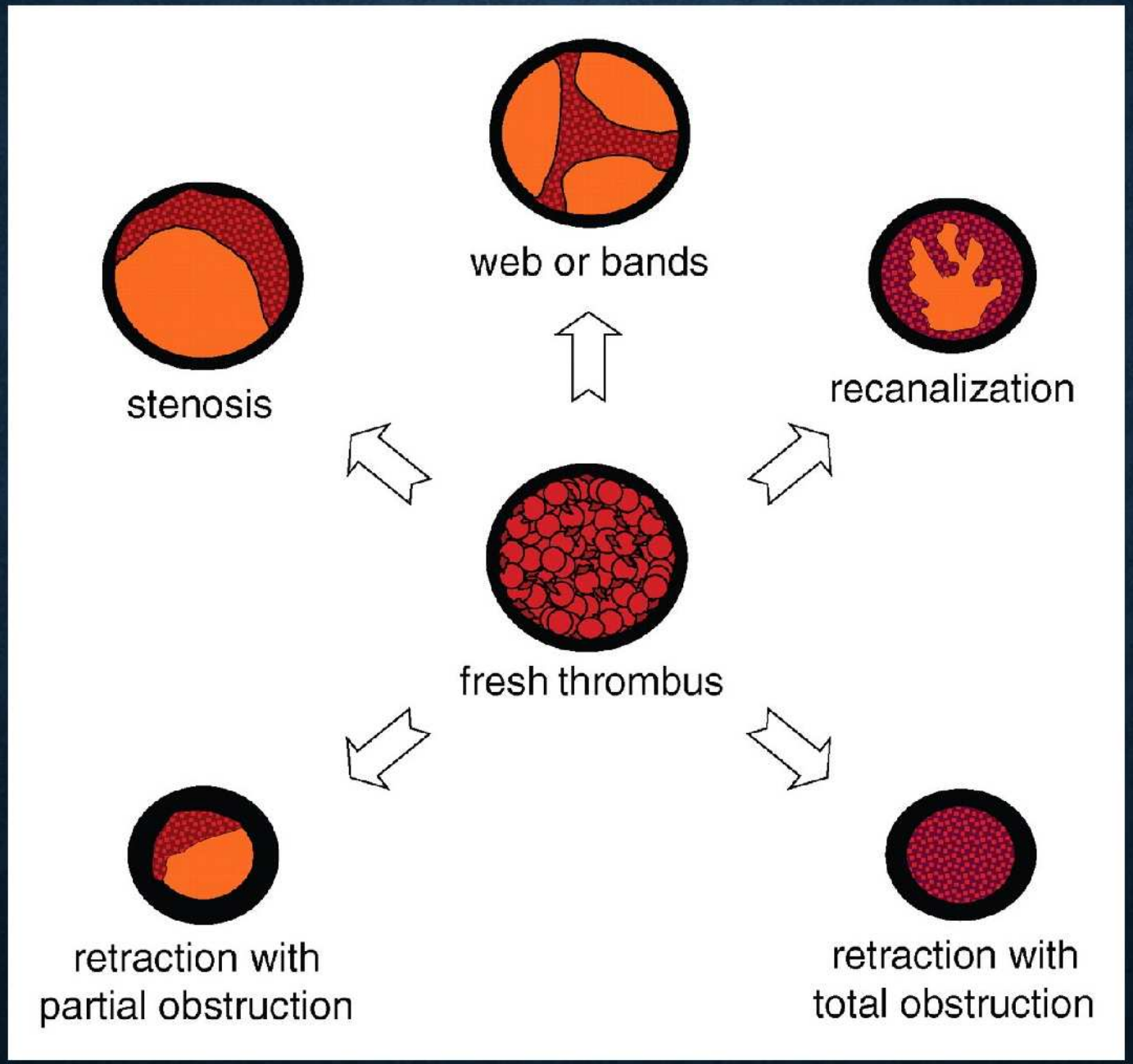

Sn140kVp
100kVp
Average
PBV
Fused image

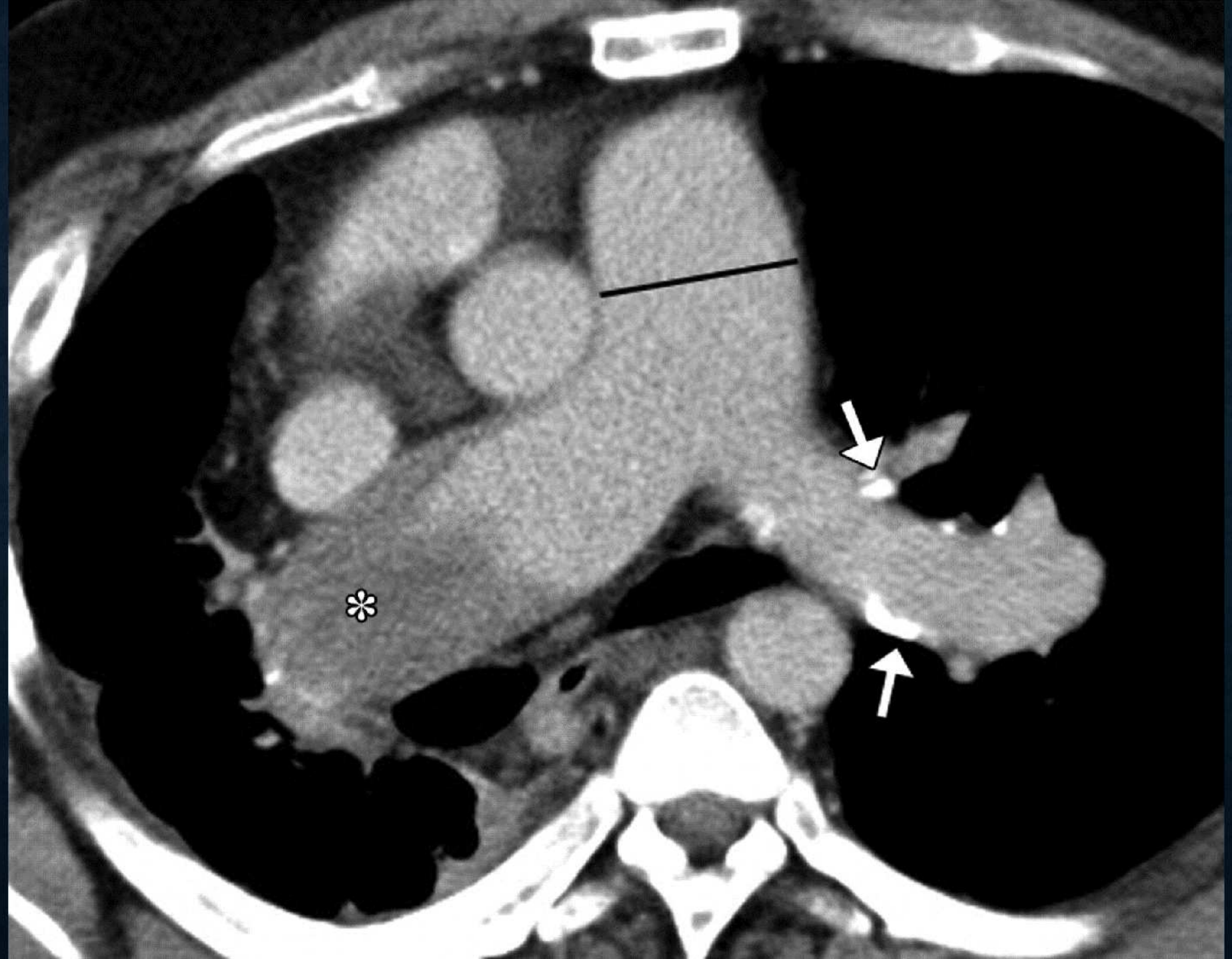
VEEL HUVITAVAT

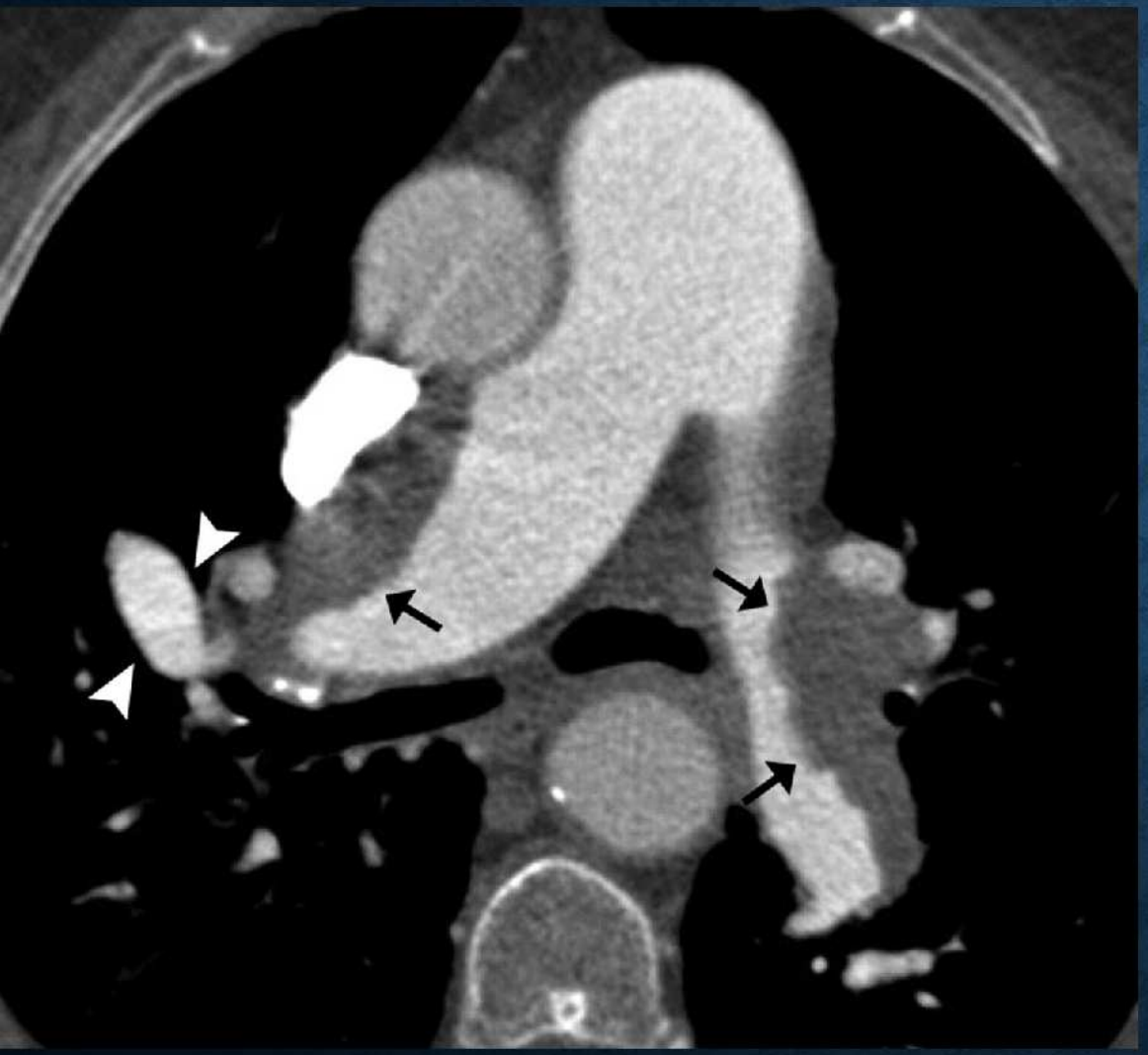
- Kahe energiaga kopsuarteri KT-angiograafia protokolli on võimalik modifitseerida
- Muuta trigger ROI kopsuarter →aort; trigger lävi kõrgema HU peale
  - Saab kontrasteerida süsteemset vereringet ehk hinnata südame vasakut poolt ja bronhiaalartereid (kliiniliselt oluline)
- Lisades EKG-sünkronisatsiooni ja gating-ut
  - Teoreetiliselt sama uuringu käigus saaks hinnata koronaarartereid
- Sisuliselt saab teha kolm-ühes uuringut

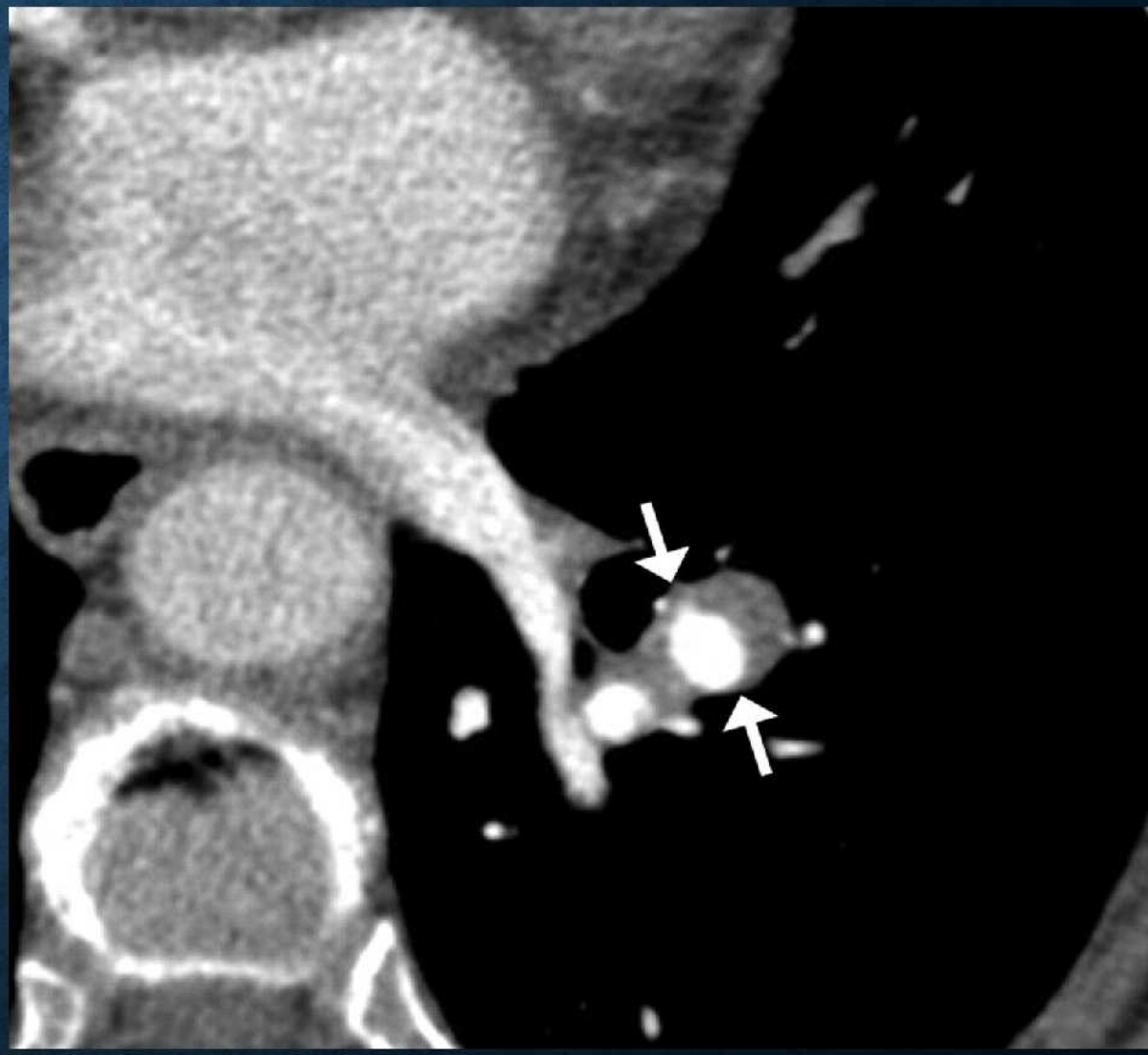
KT-UURINGUL VASKULAARSED TUNNUSED

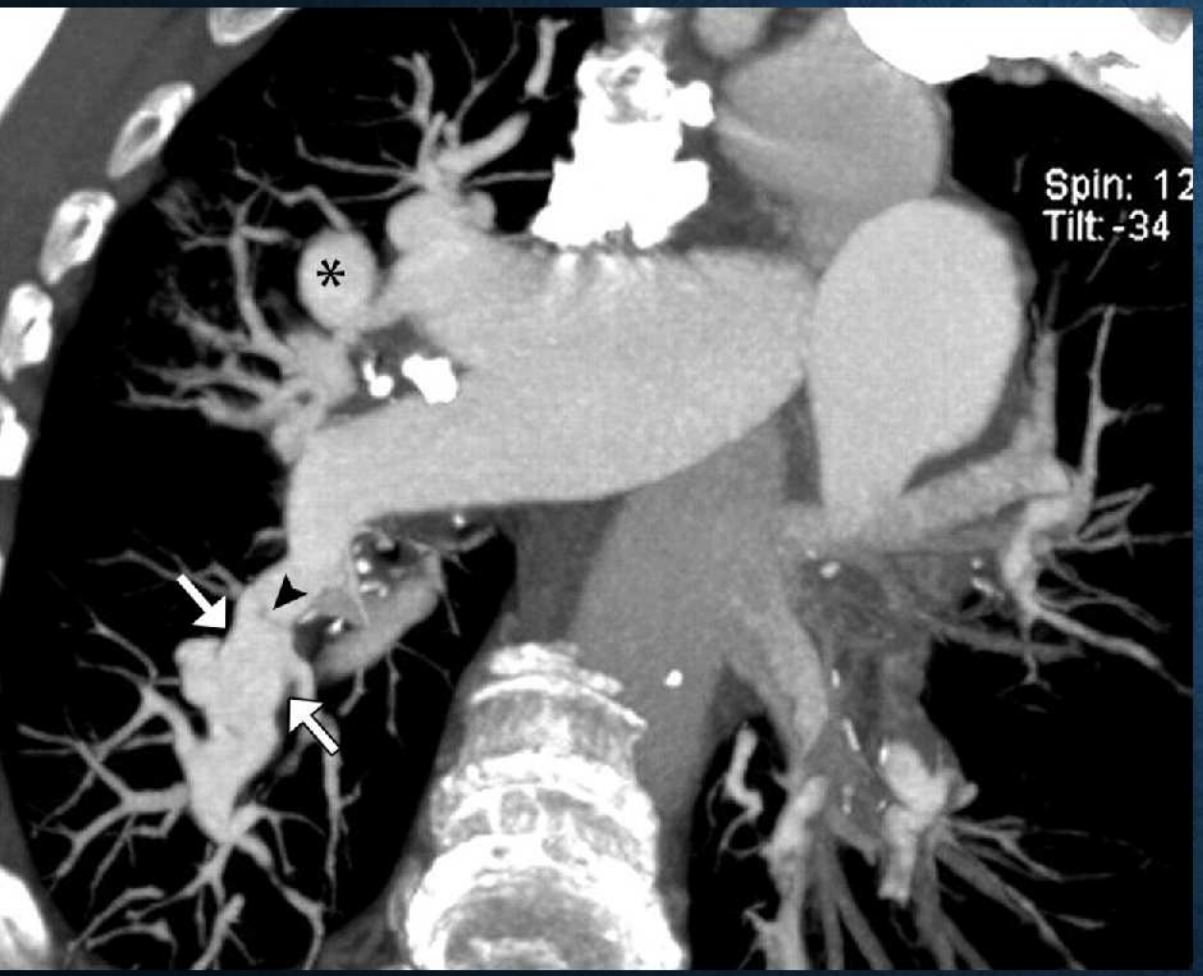
- Täielik obstruktsioon?
- Miiteoklusiivsed täitedefektid
  - Ekstsentrilised ehk seinapidised (vanad) vs kontsentrilised (pigem värsked)
- Arterid (v.a. peatüvi) kitsenenud vs ägedas faasis läbimõõt muutusteta või laiem, võimalik poststenootiline laienemine
- Pulmonaalhüpertensioonile viited
  - Laienenud kopsuarteri peatüvi (üle 29 mm aksiaalkihil laiemas kohas), distaalsemal iseloomulik pigem asümmeetriline laienemine (vs sümmeetriline mitte-CTEPH juhtudel)
  - Kopsuarteri peatüve läbimõõt suurem kui samas kihis üleneva aordi läbimõõt
  - Parema vatsakese laienemine, interventrikulaarseptumi nihe vasakule (nn McConnell-i tunnus), RV vabaseina hüpertroofia
- Kollateraalid
  - Bronhiaalarterite laienemine (N proksimaalsel kuni 1,5 mm)

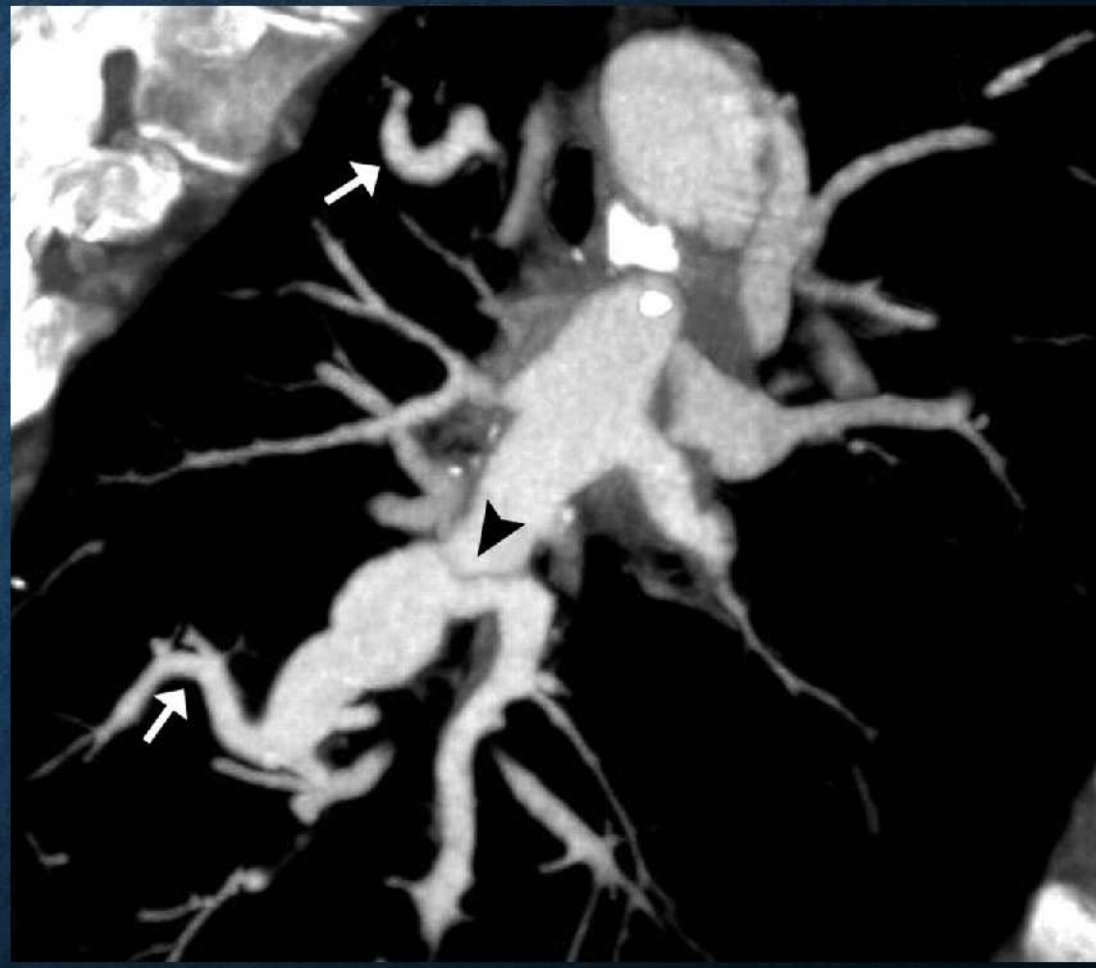

web or bands
stenosis
recanalization
fresh thrombus
retraction with
partial obstruction
retraction with
total obstruction

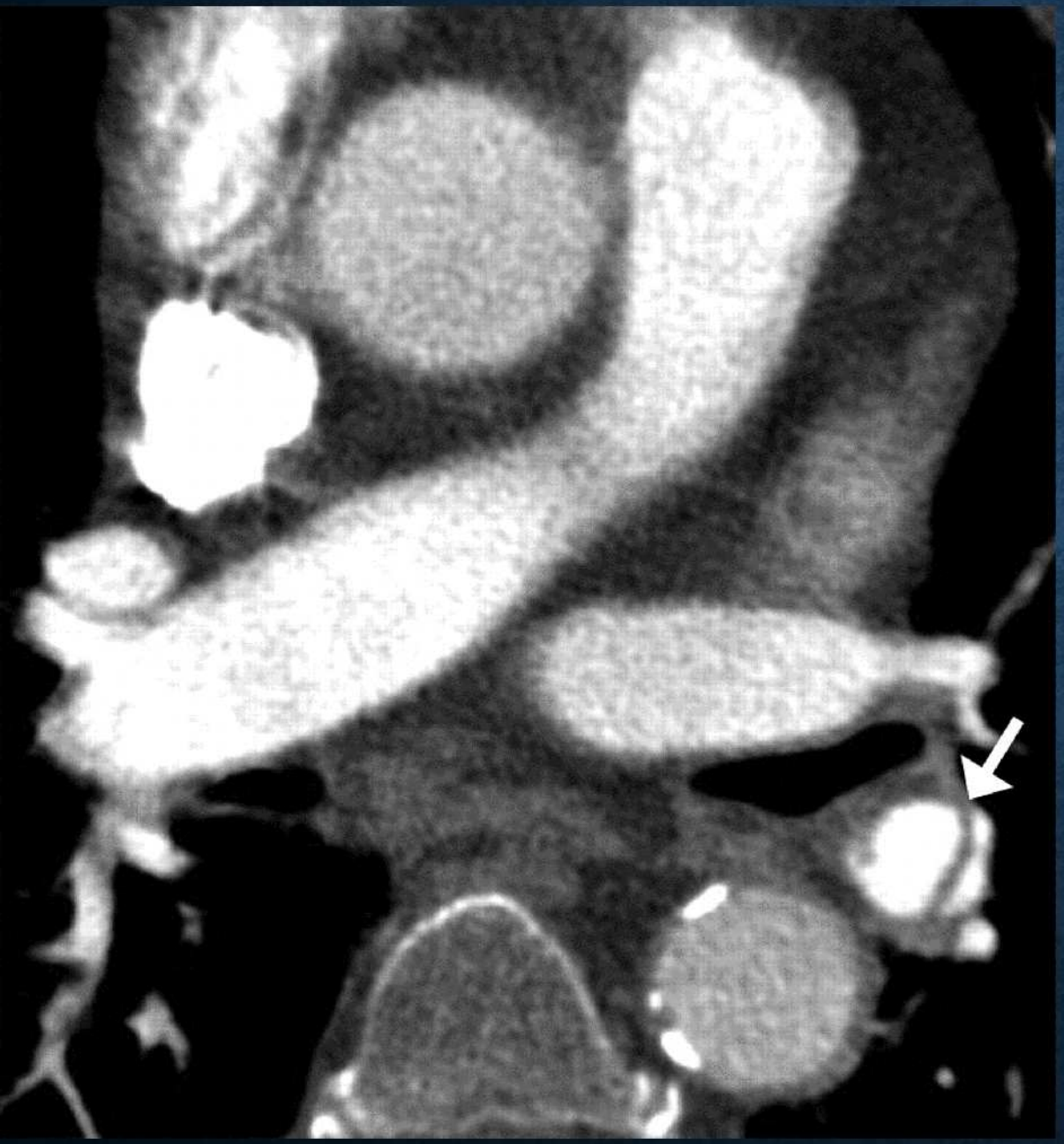


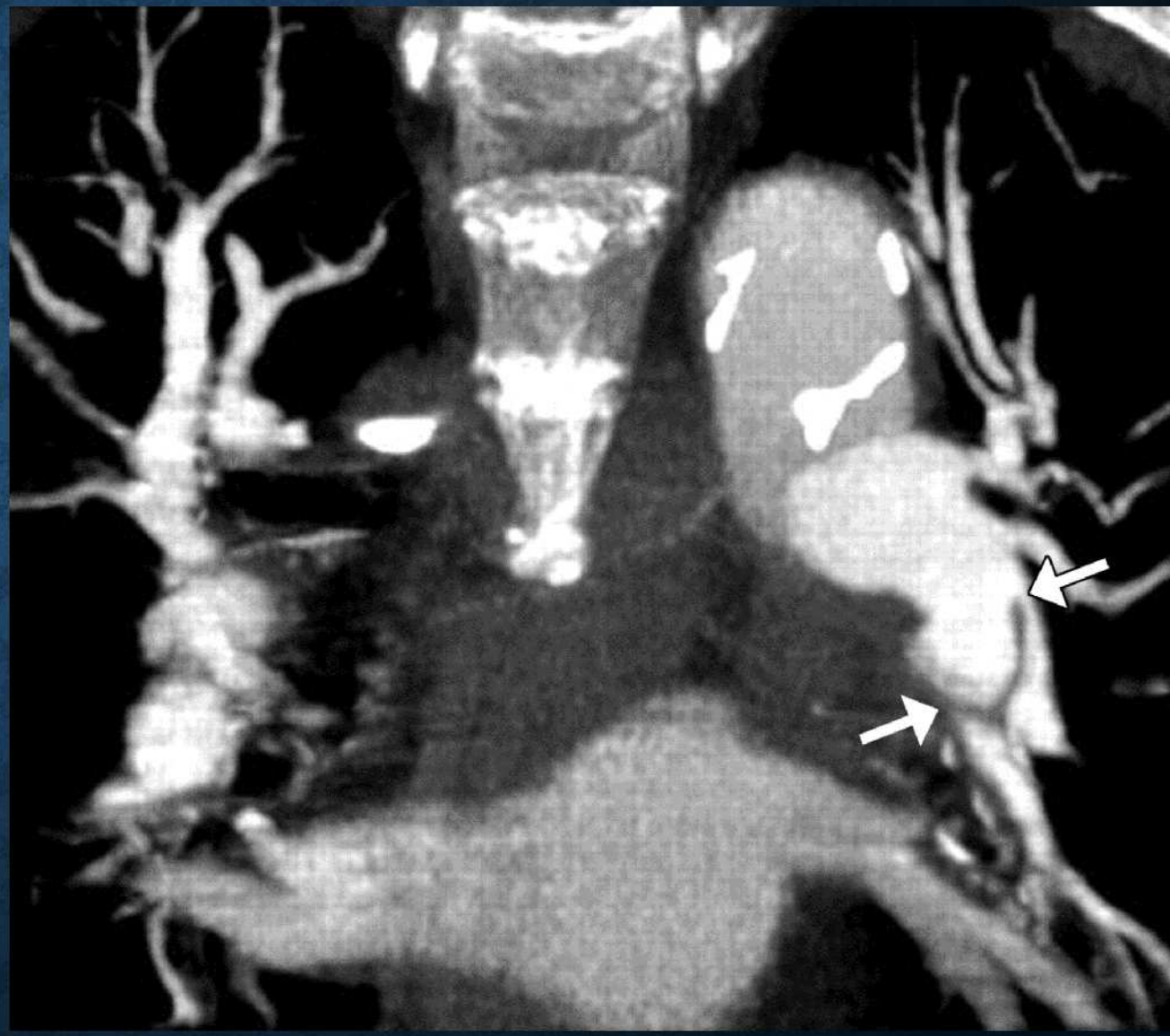


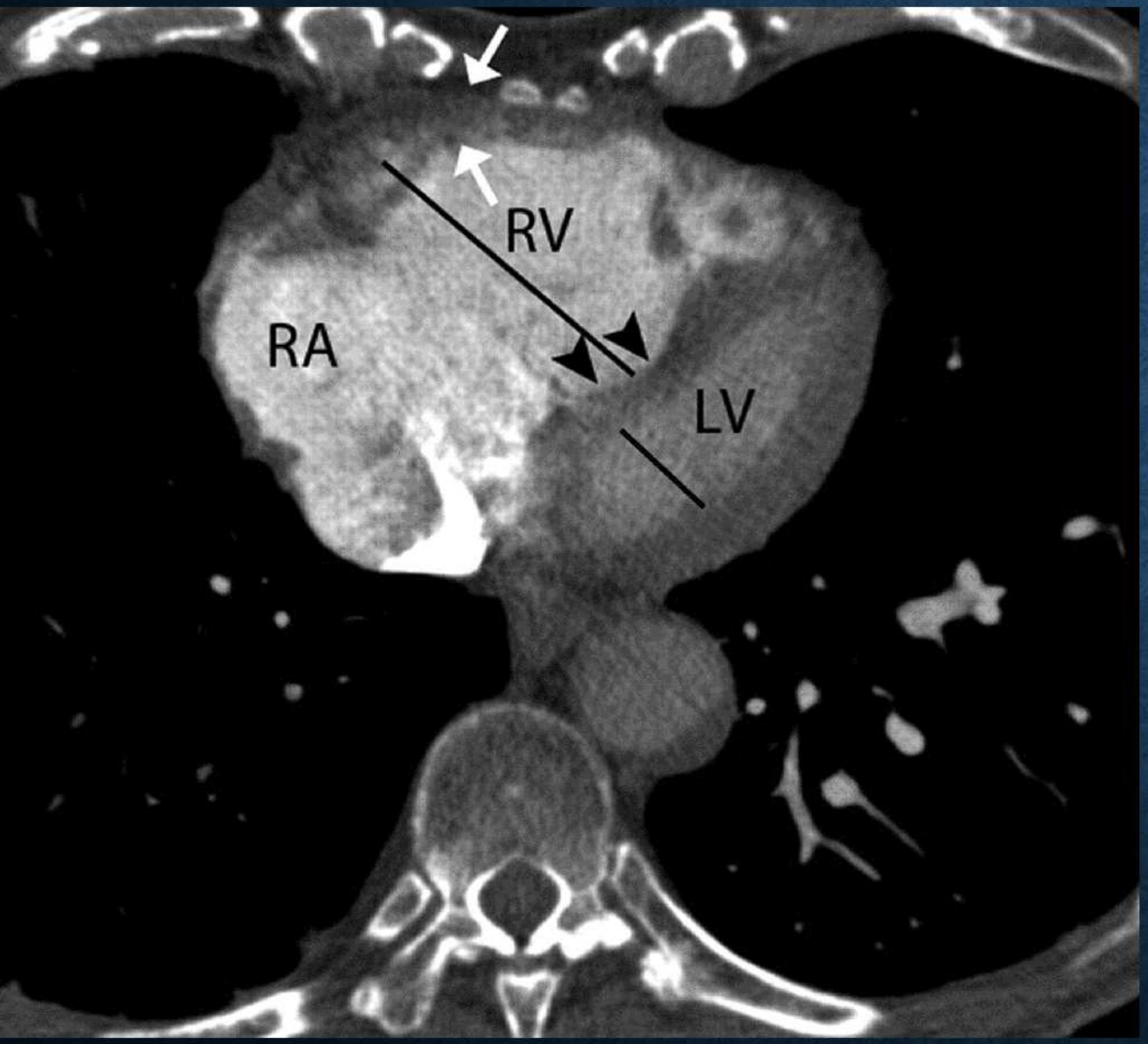


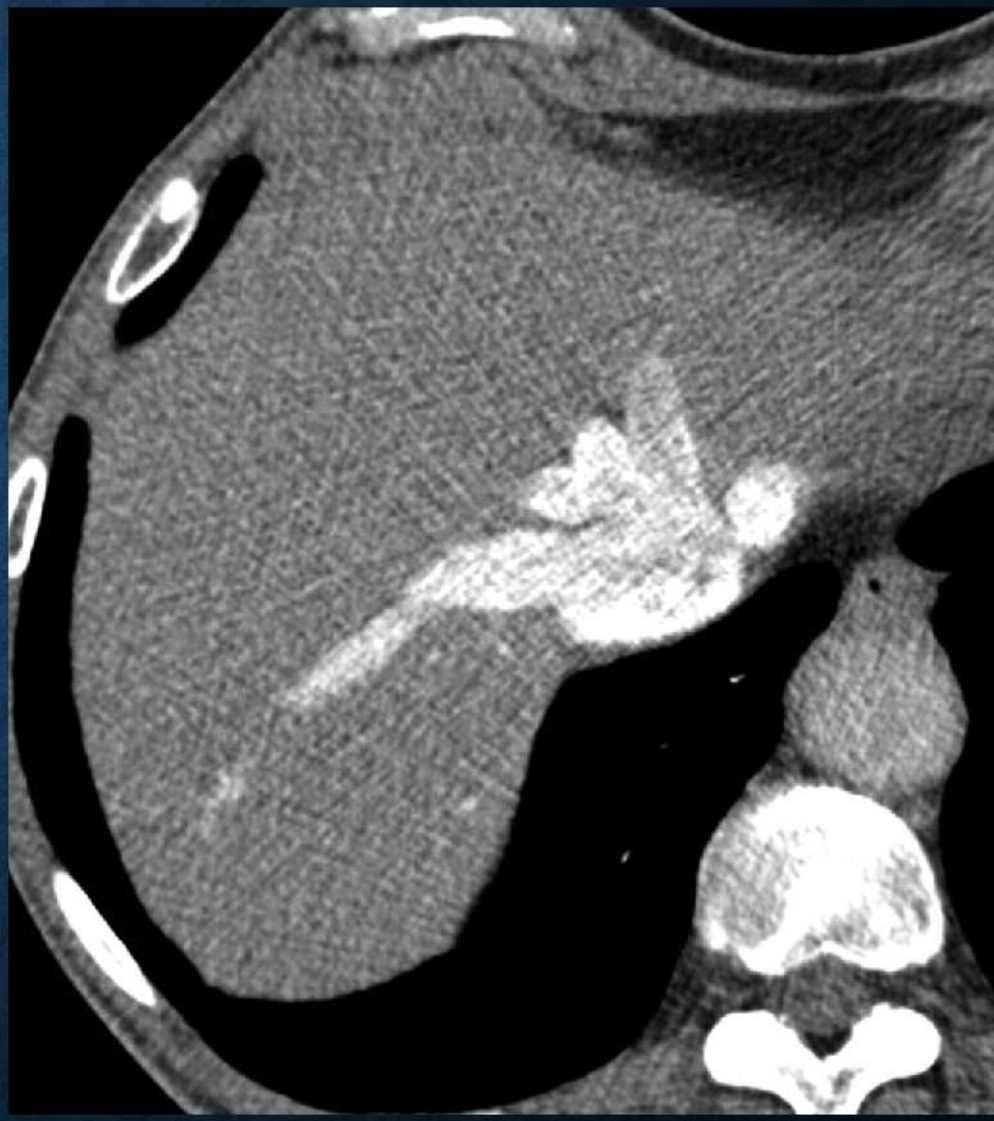

Spin: 12
Tilt: -34

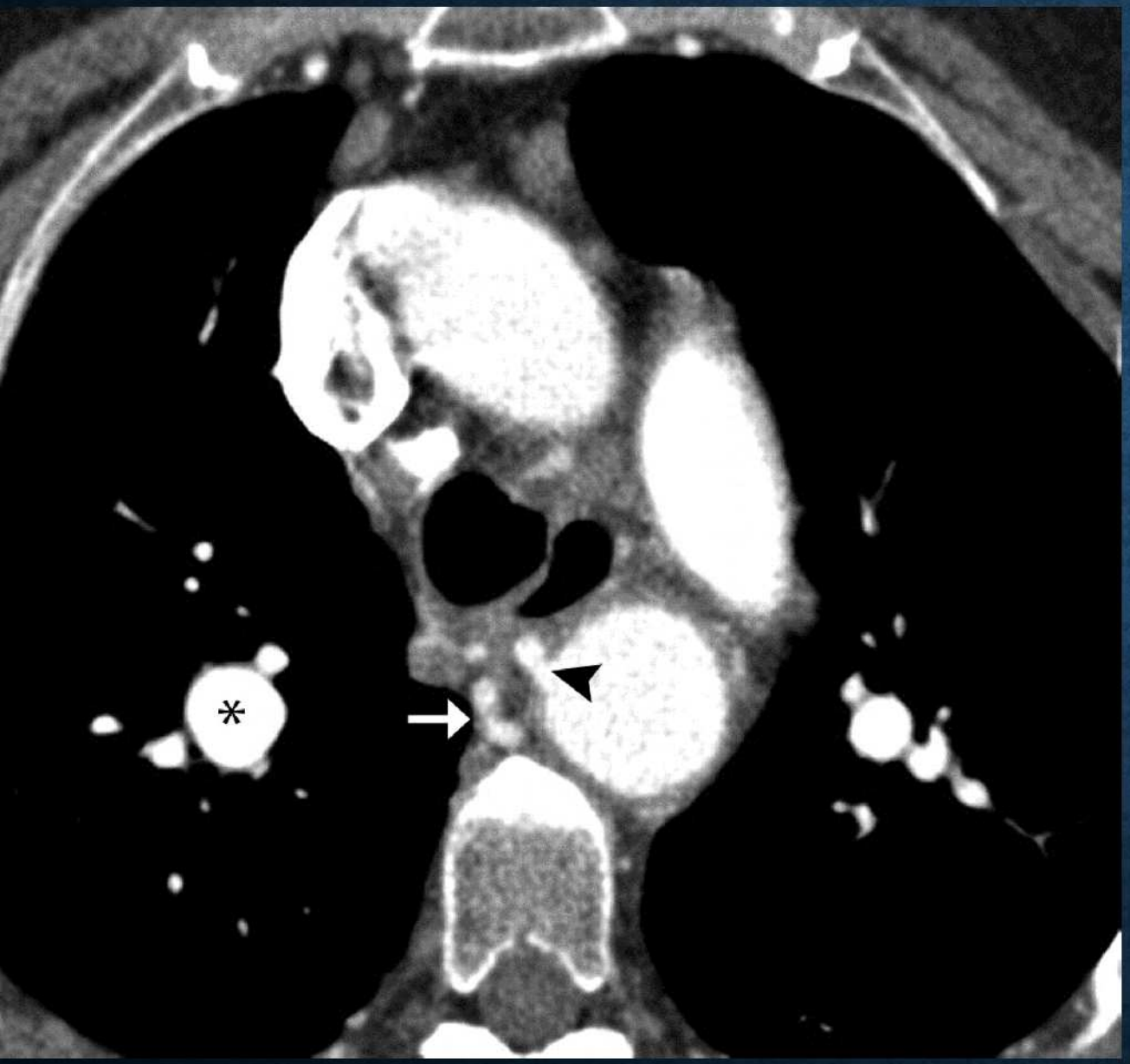


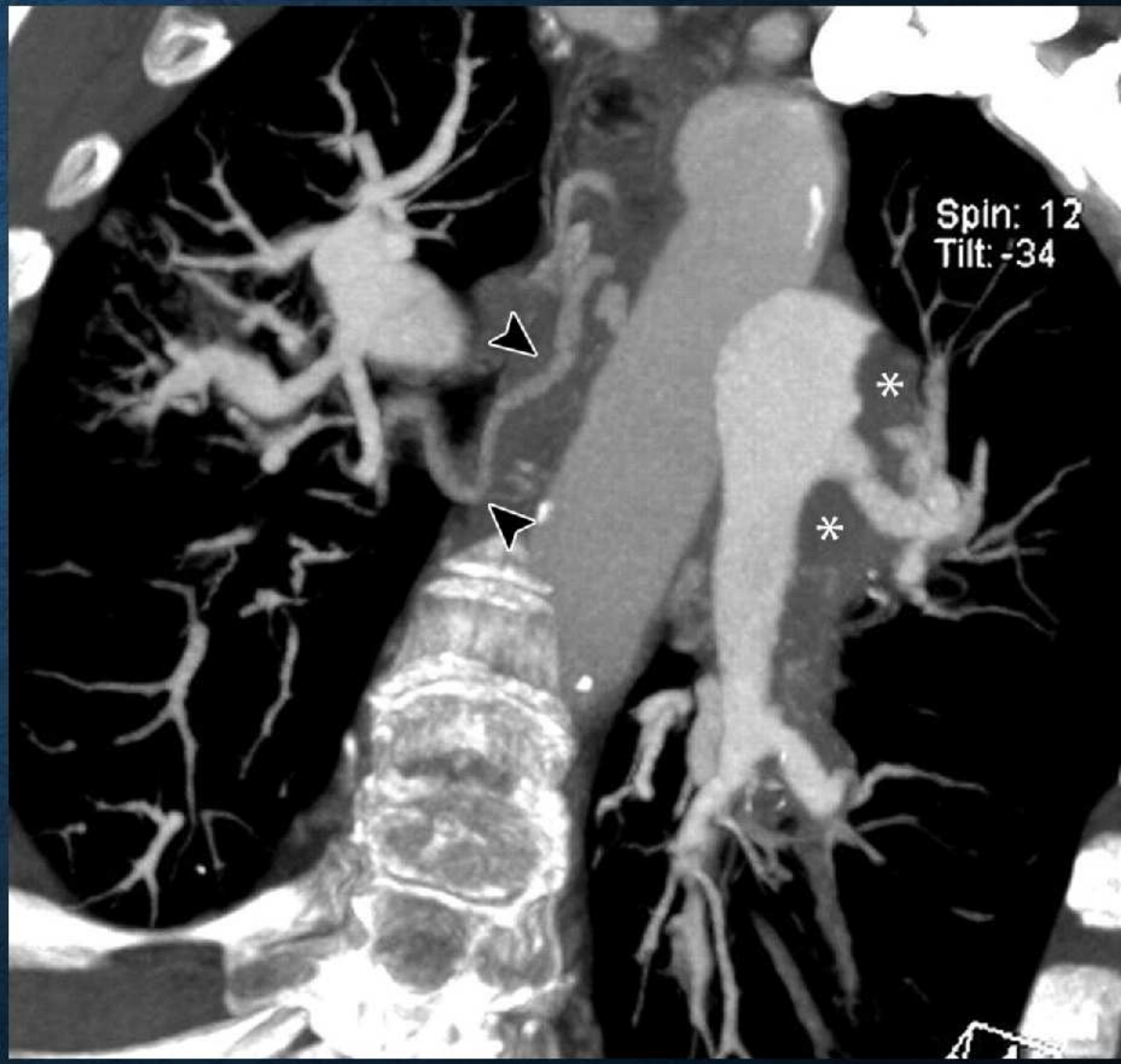





RV
RA
LV

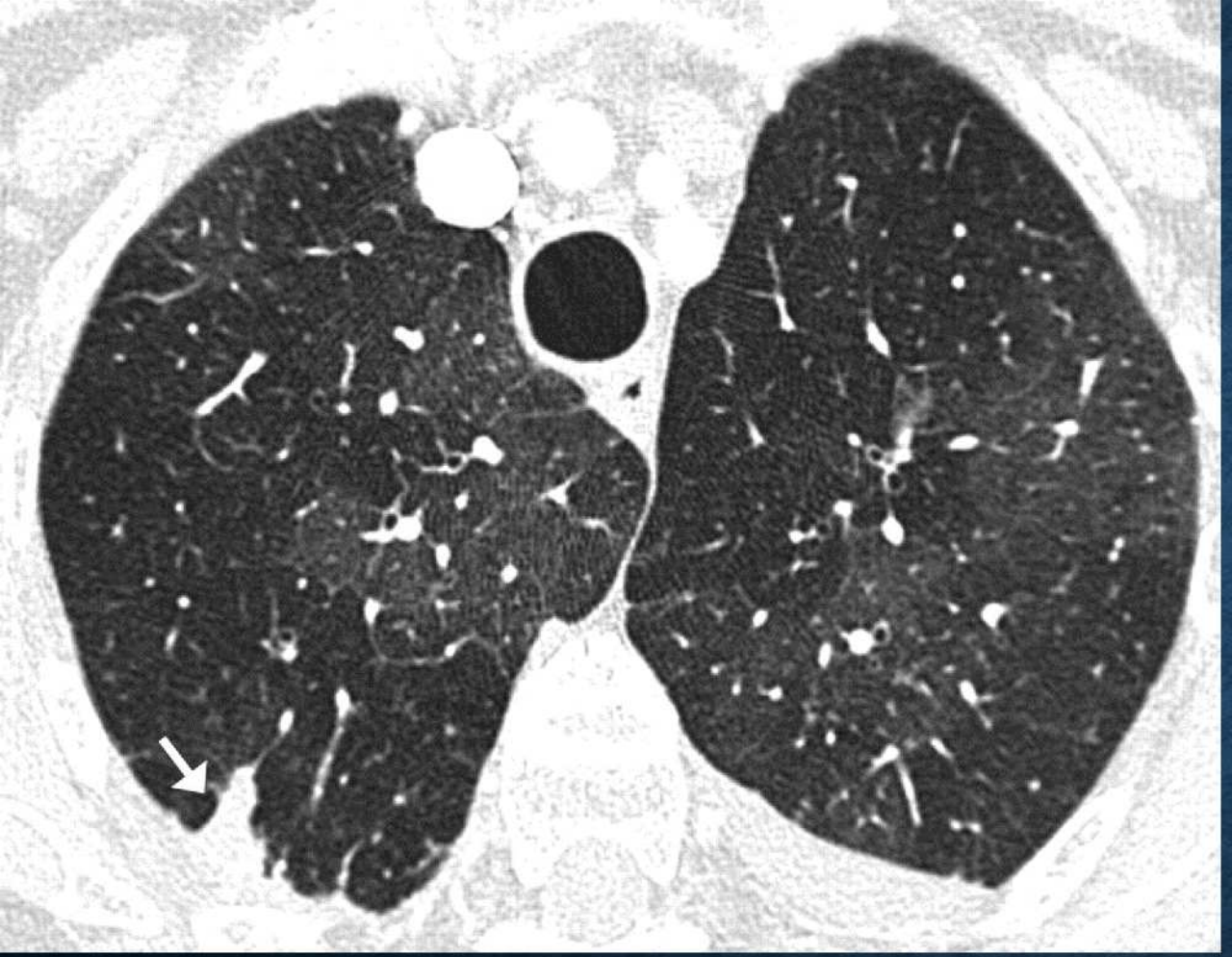


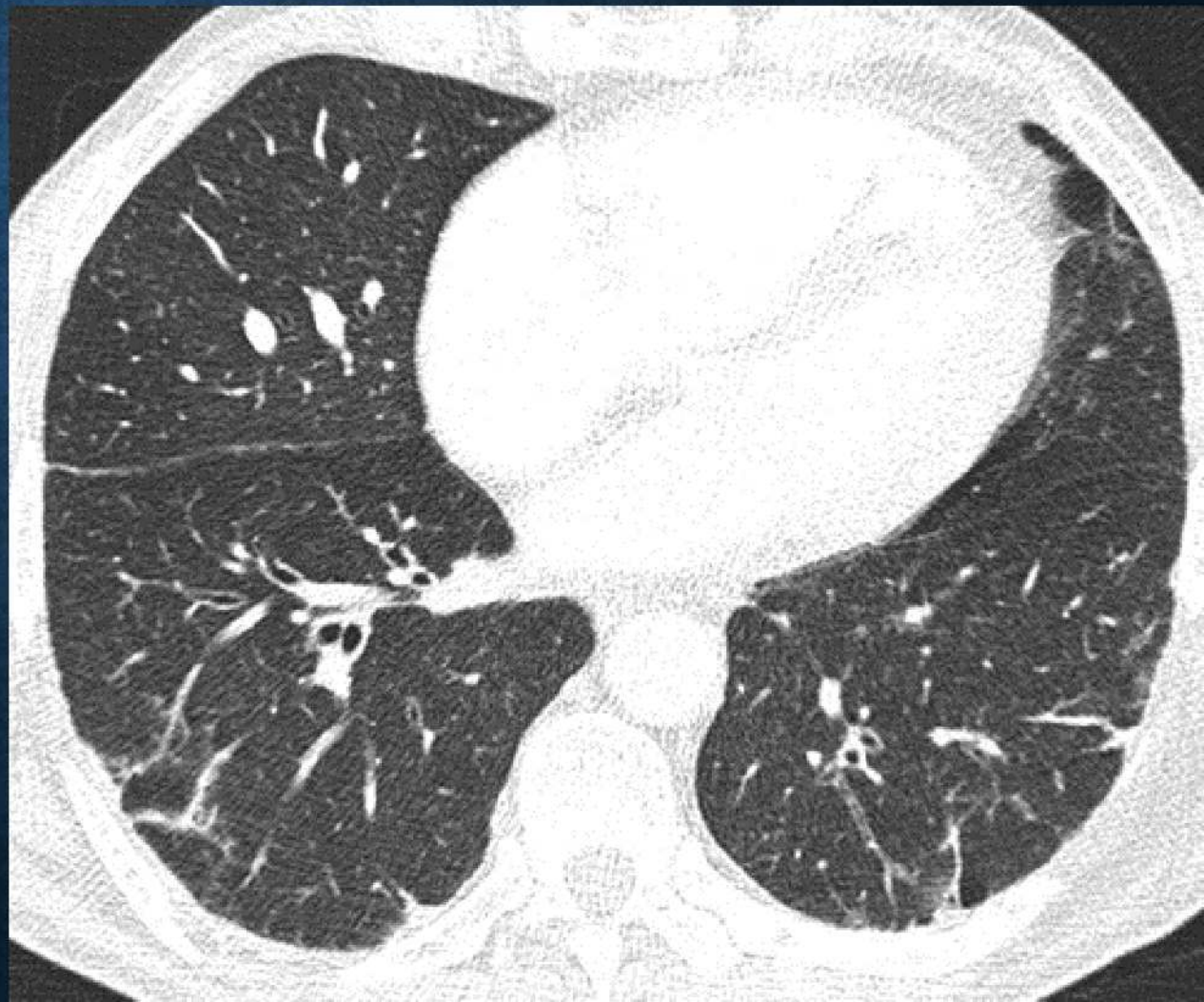


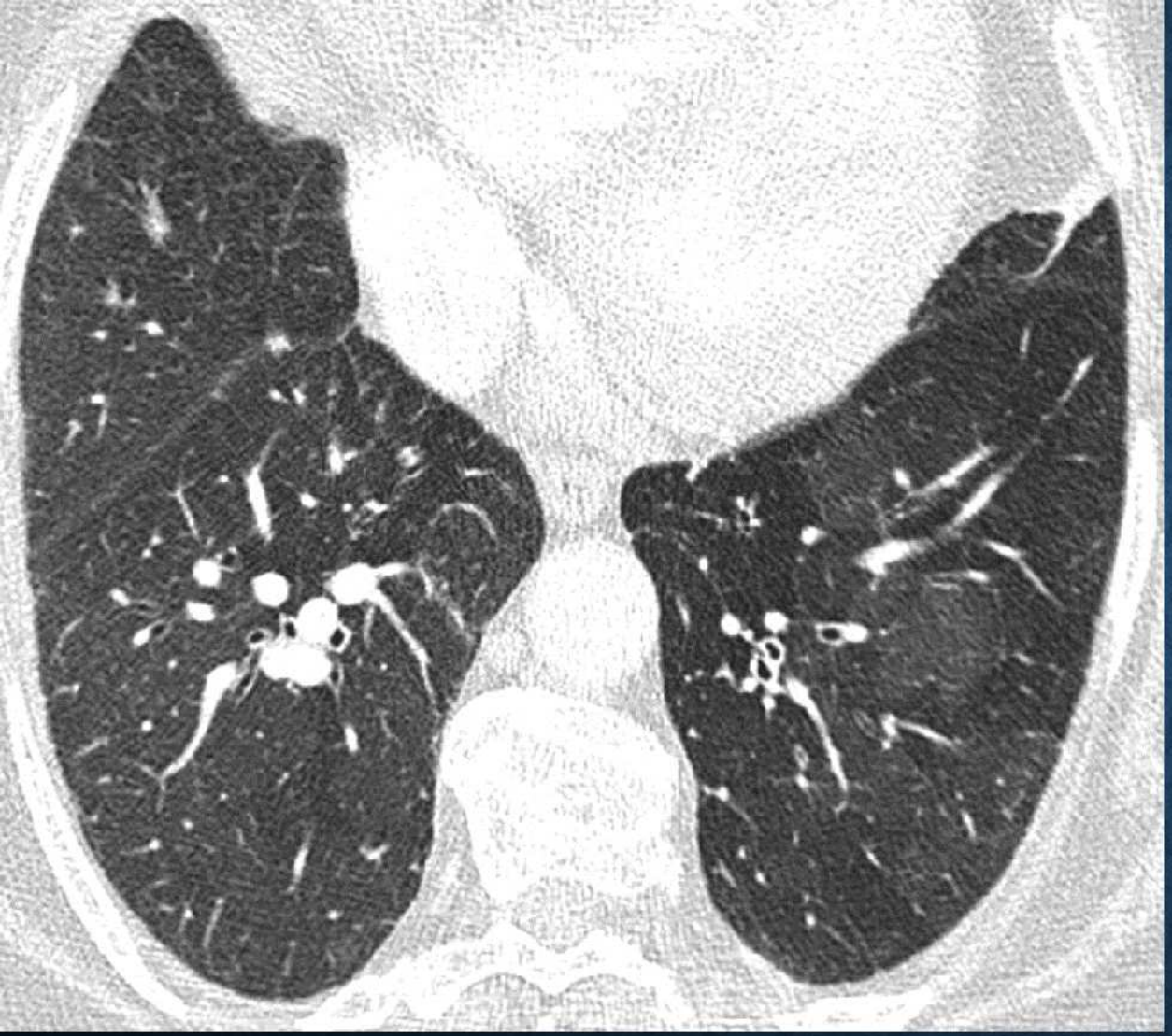

Spin: 12
Tilt: -34

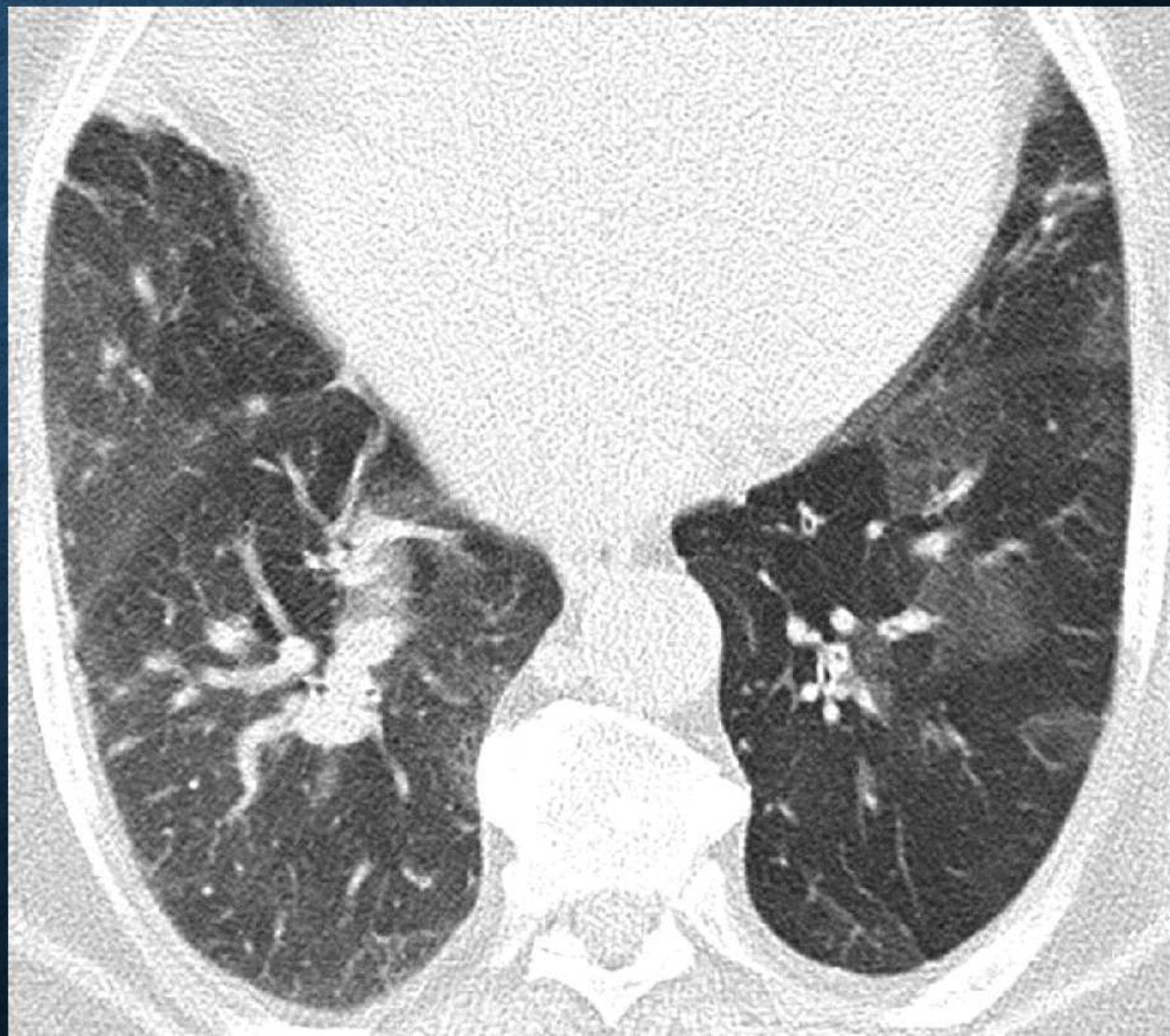
PARENHÜMATOOSSED TUNNUSED

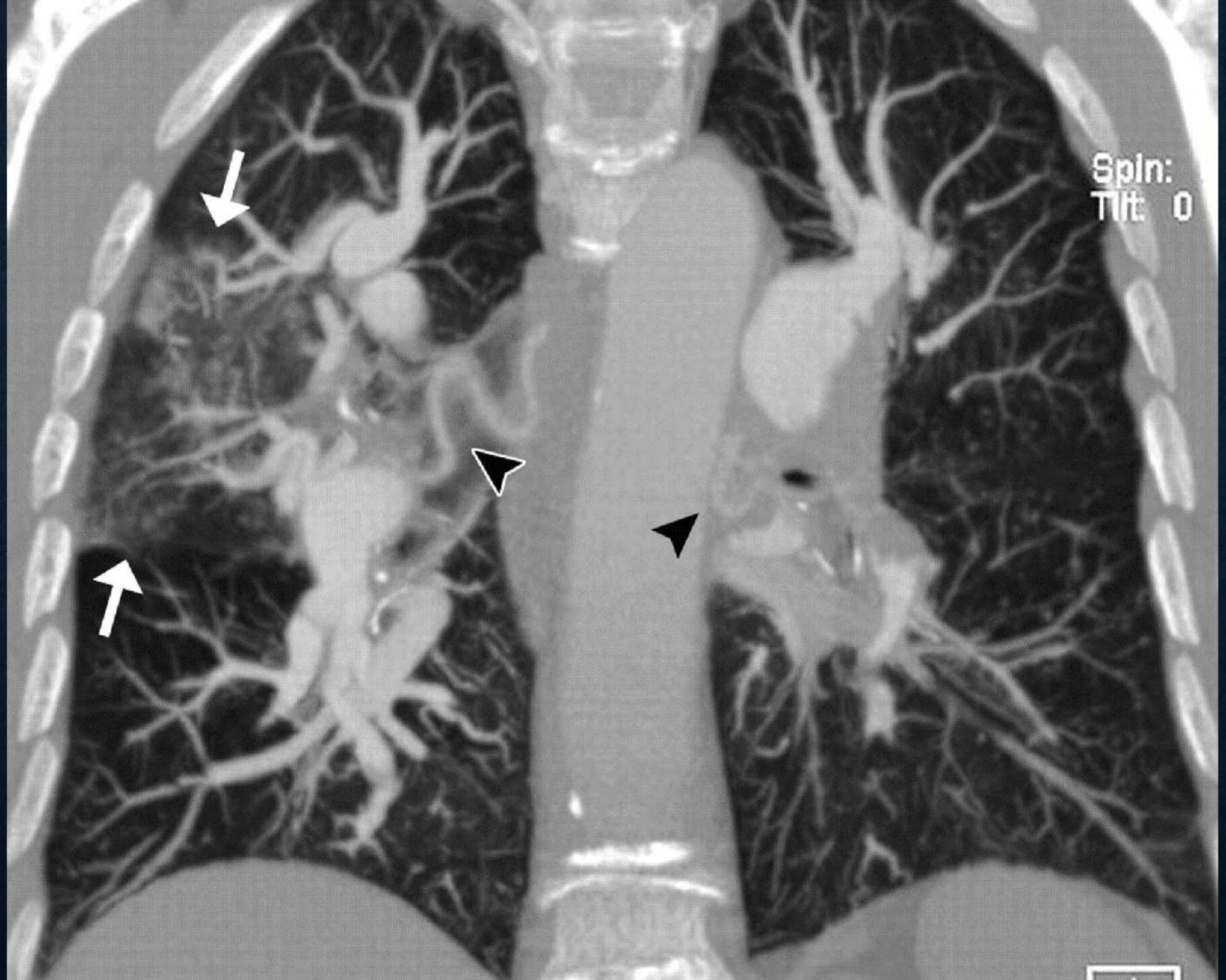
- Vanadest kopsuinfarktidest kiiljad perifeersed fibrootilised alad
- Mosaiikperfusiooni muster
- Mattklaas-muutused süsteemsetest kollateraalidest





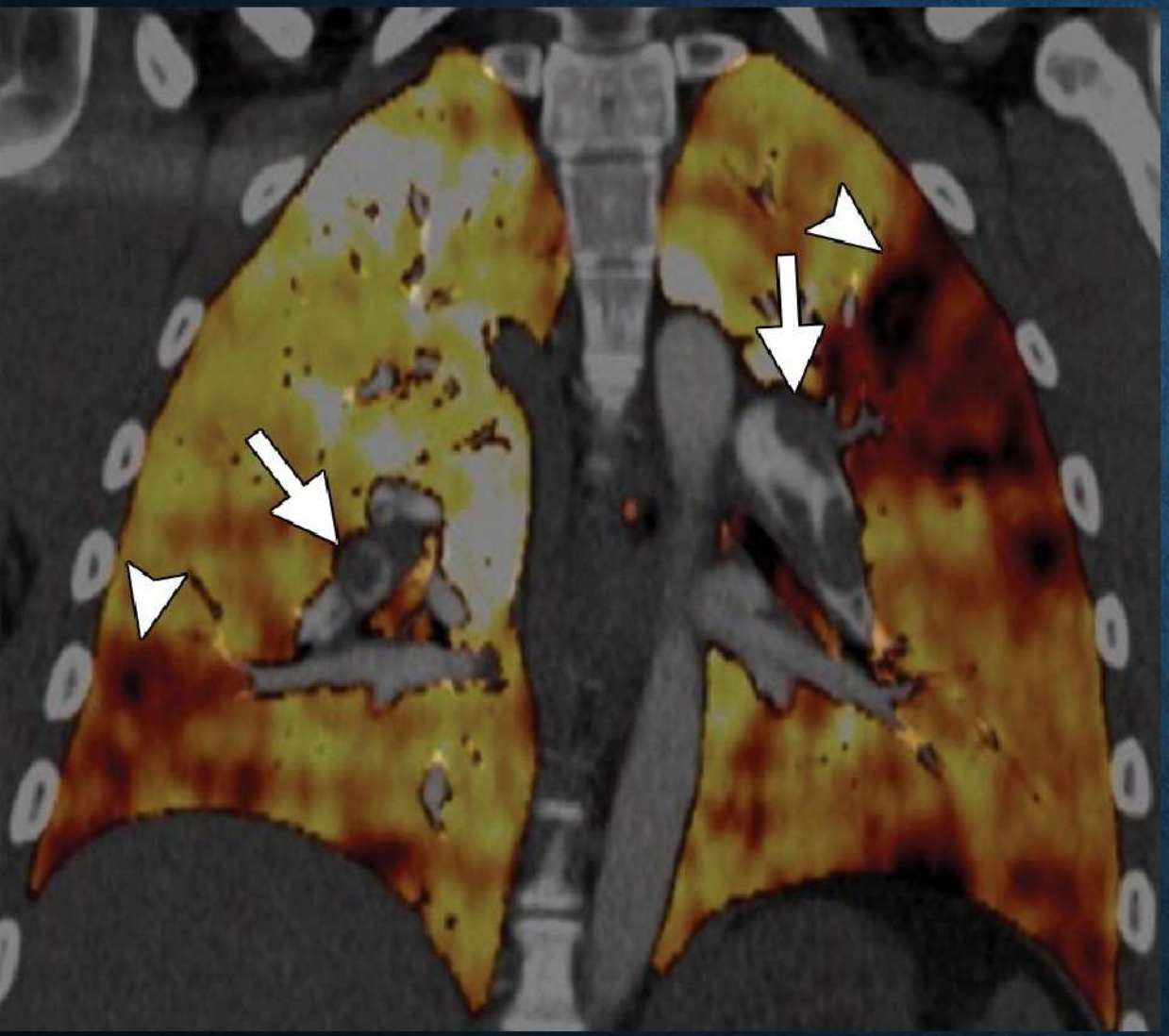


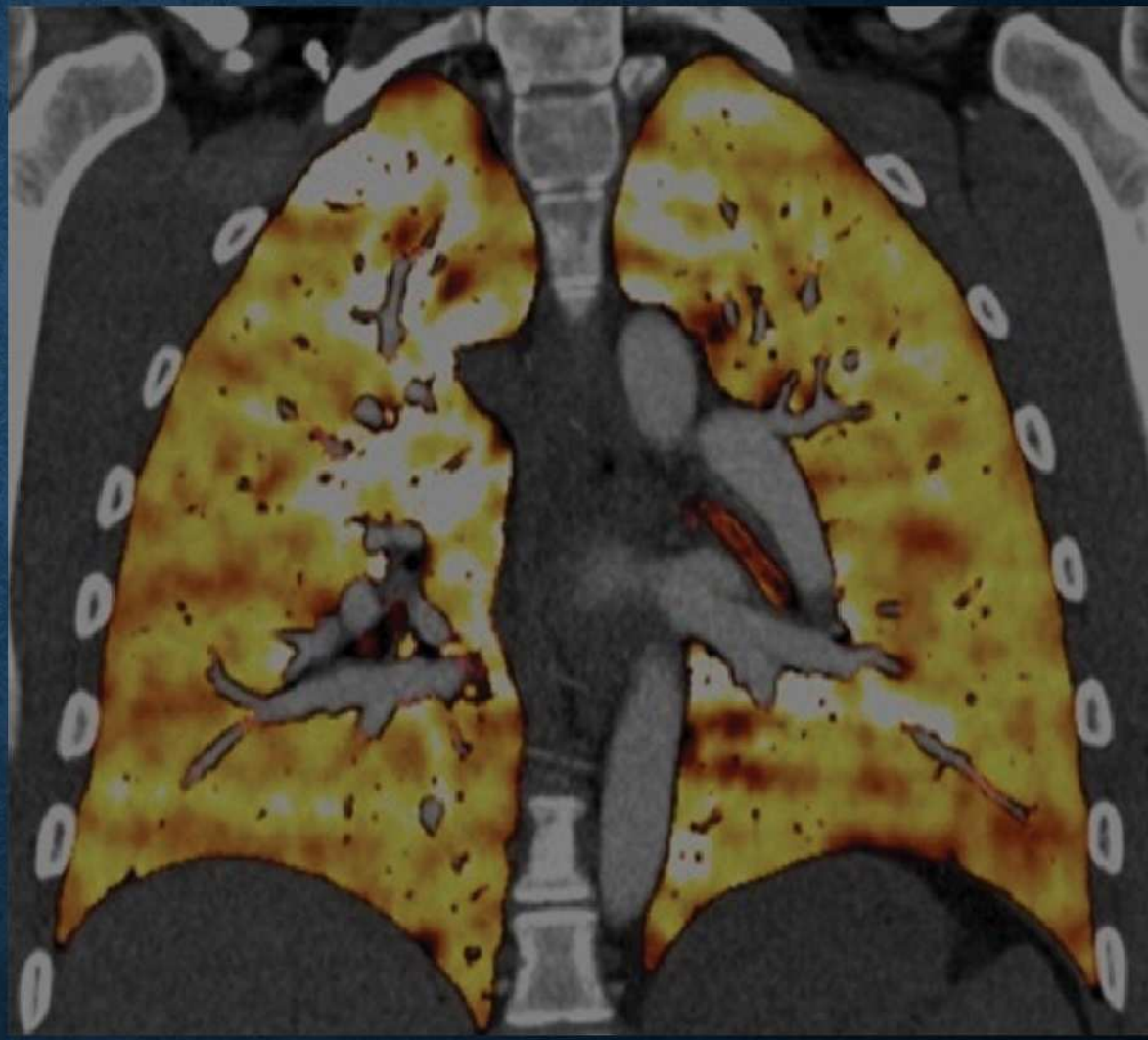


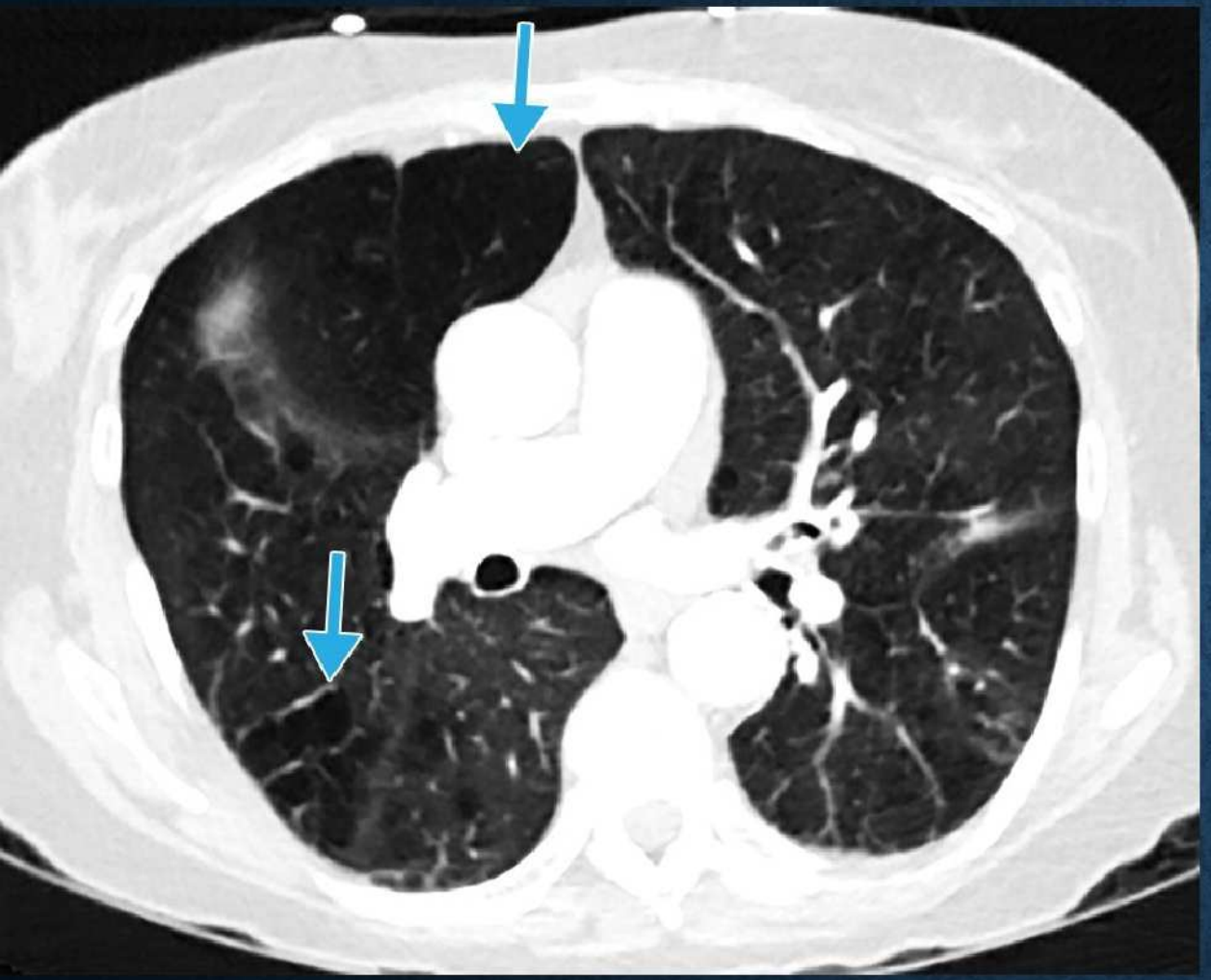

Spin:
Tilt: 0

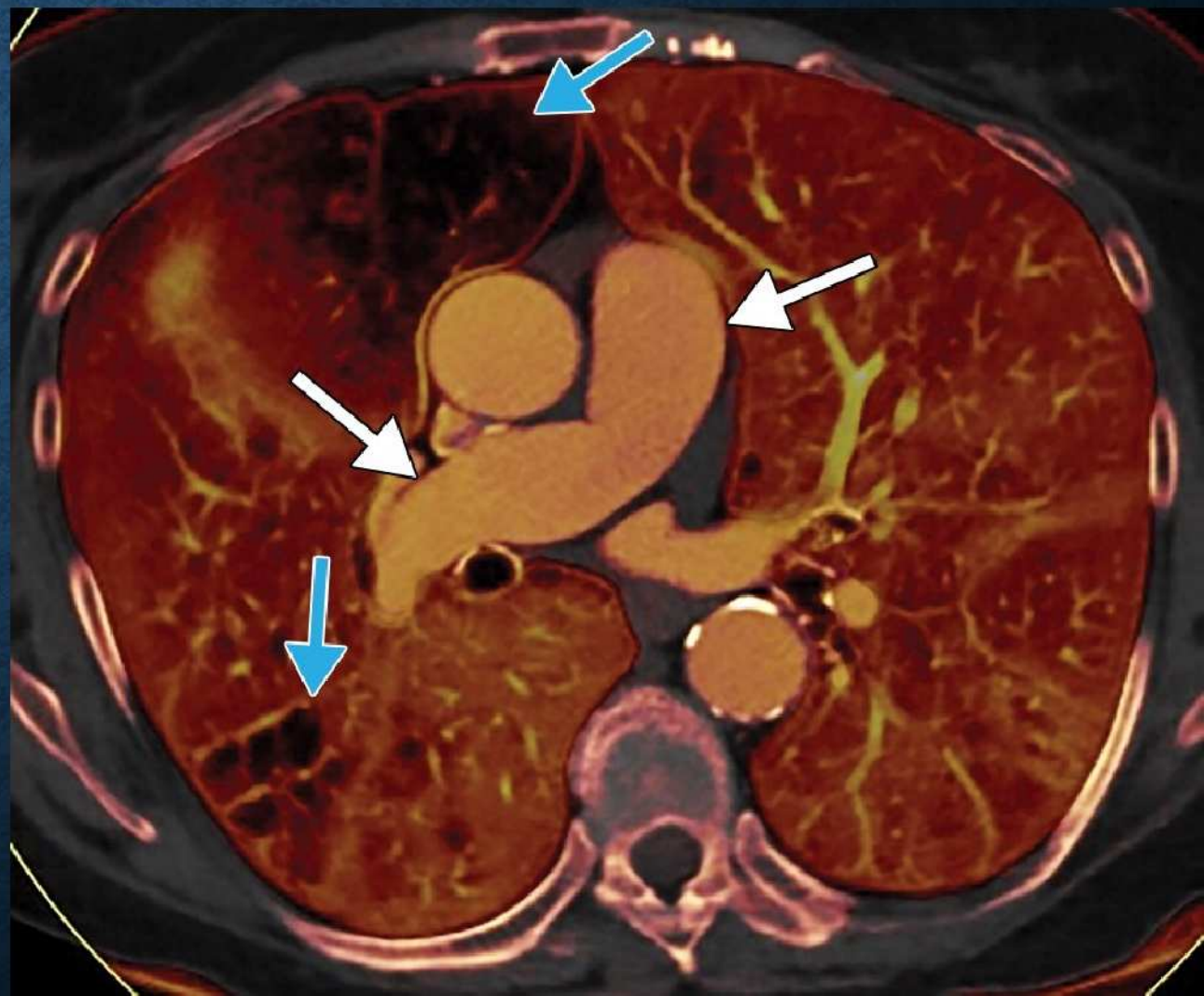
DECT JA PBV-KAARDID

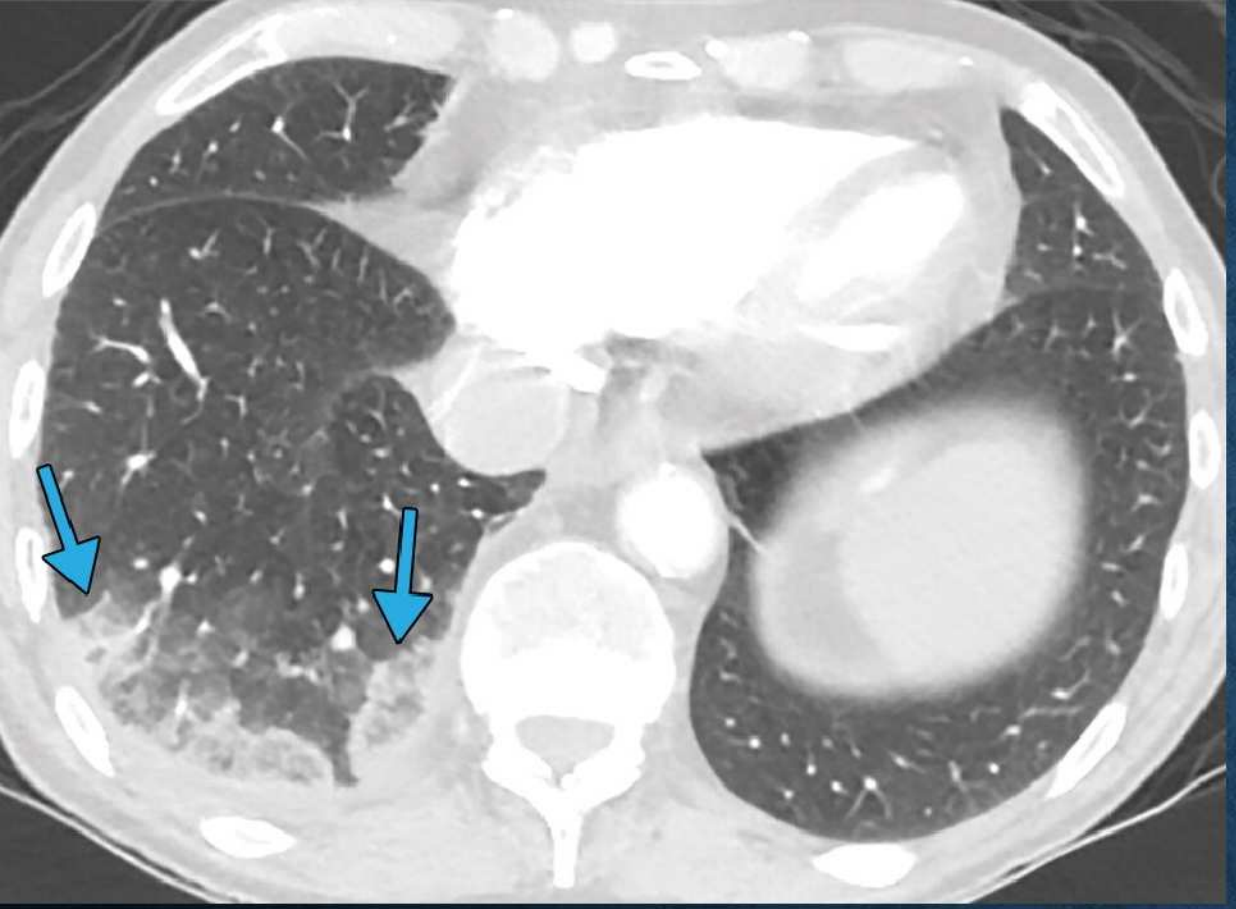
- Kopsuarteri muutuse vastandamine langenud regionaalsele perfusioonile
- Hüpoperfusiooni ala PBV kaardil võib olla suurem kui kopsu kahjustus „segaenergiaga“ seerial
- Oluline hinnata ka ilma PBV-kaardita uuring
  - Muud põhjused regionaalse perfusiooni langusele nt konsolidatsioonid või emfüseem

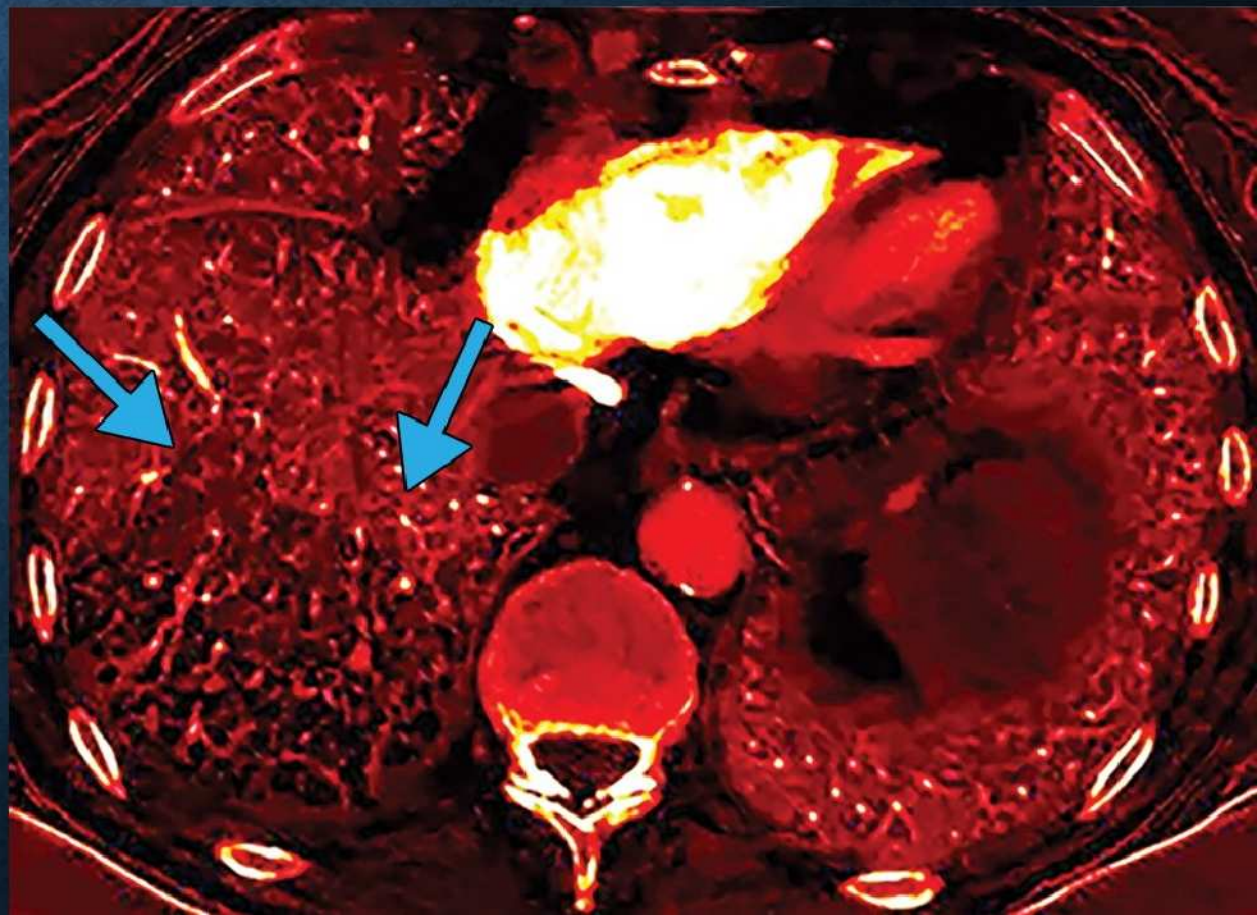




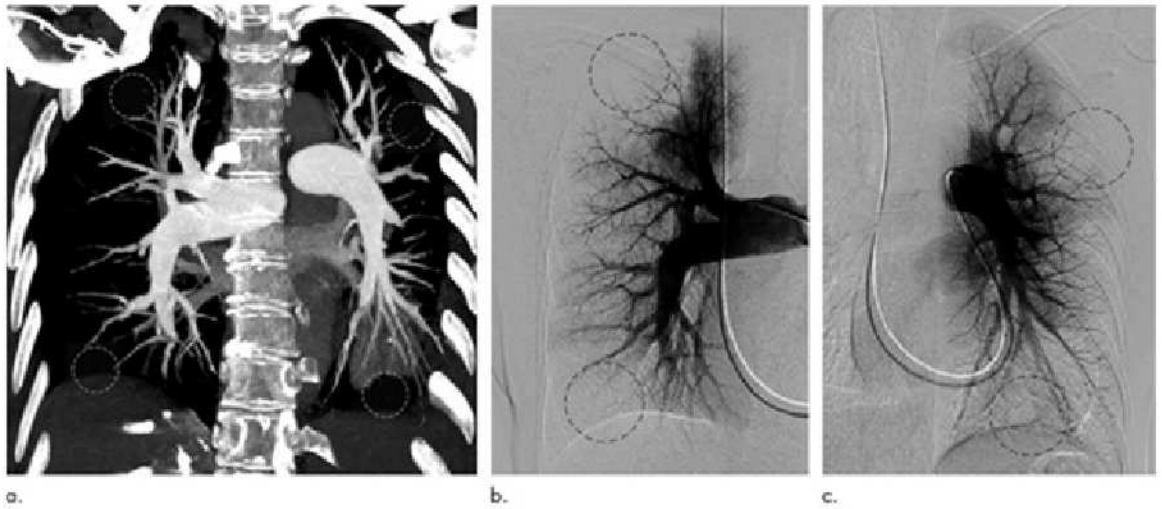








Imaging of Pulmonary Hypertension in Adults: A Position Paper from the Fleischner Society

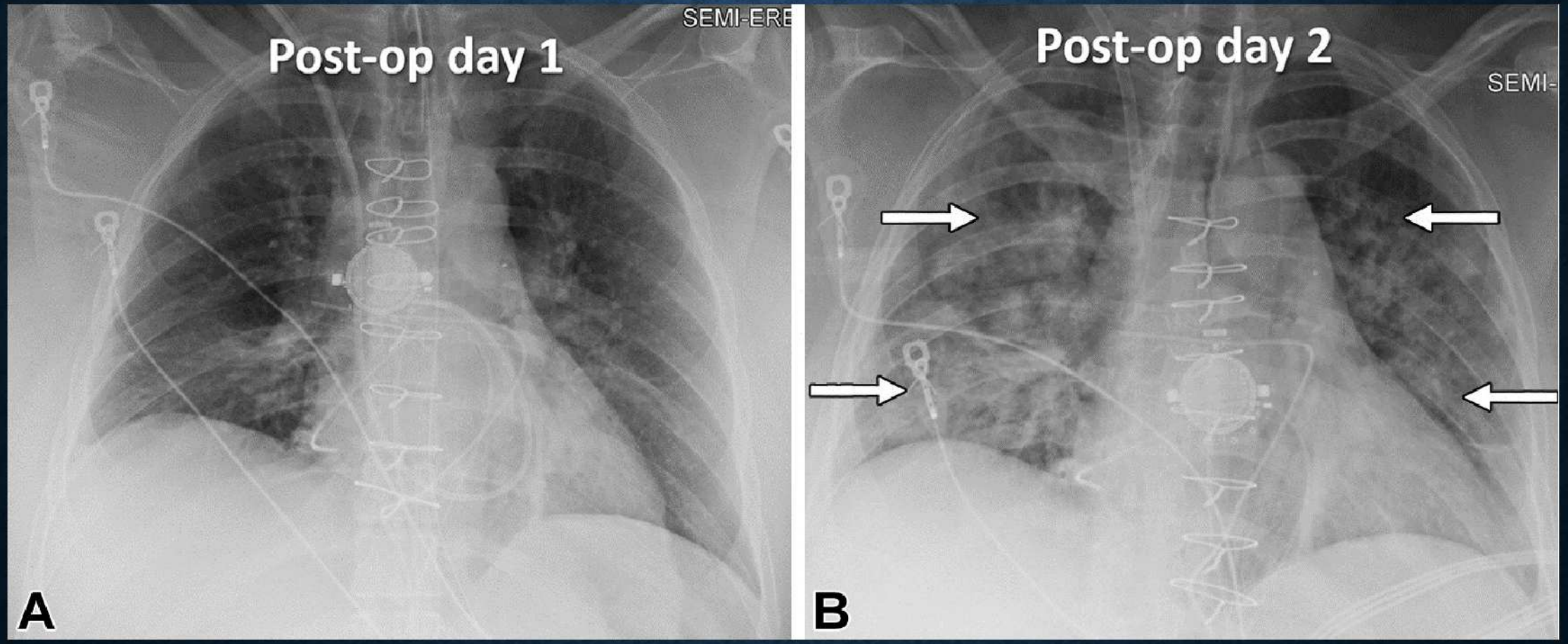


(a) CTA and **(b)** right and **(c)** left pulmonary angiograms show vessel pruning and poor distal perfusion (dotted circles) on CTA and corresponding perfusion defects (dotted circles) on the angiograms.

- Pulmonary artery diameter is insufficient as a standalone criterion for pulmonary hypertension (PH).
- V/Q scan is the recommended examination in patients with PH to rule out chronic thromboembolic PH.
- Single-energy CT provides information on PH etiology and plays important role in the diagnostic strategy.
- Dual-energy CT combines morphologic information with lung perfusion (ie, iodine maps) and can potentially increase CT diagnostic capabilities.
- Cardiac MRI has become the reference standard to determine right ventricular function in patients with PH.

Remy-Jardin M et al. Published Online: January 5, 2021
https://doi.org/10.1148/radiol.2020203108

Radiology


Post-op day 1
SEMI-ERE
A
Post-op day 2
SEMI-
B

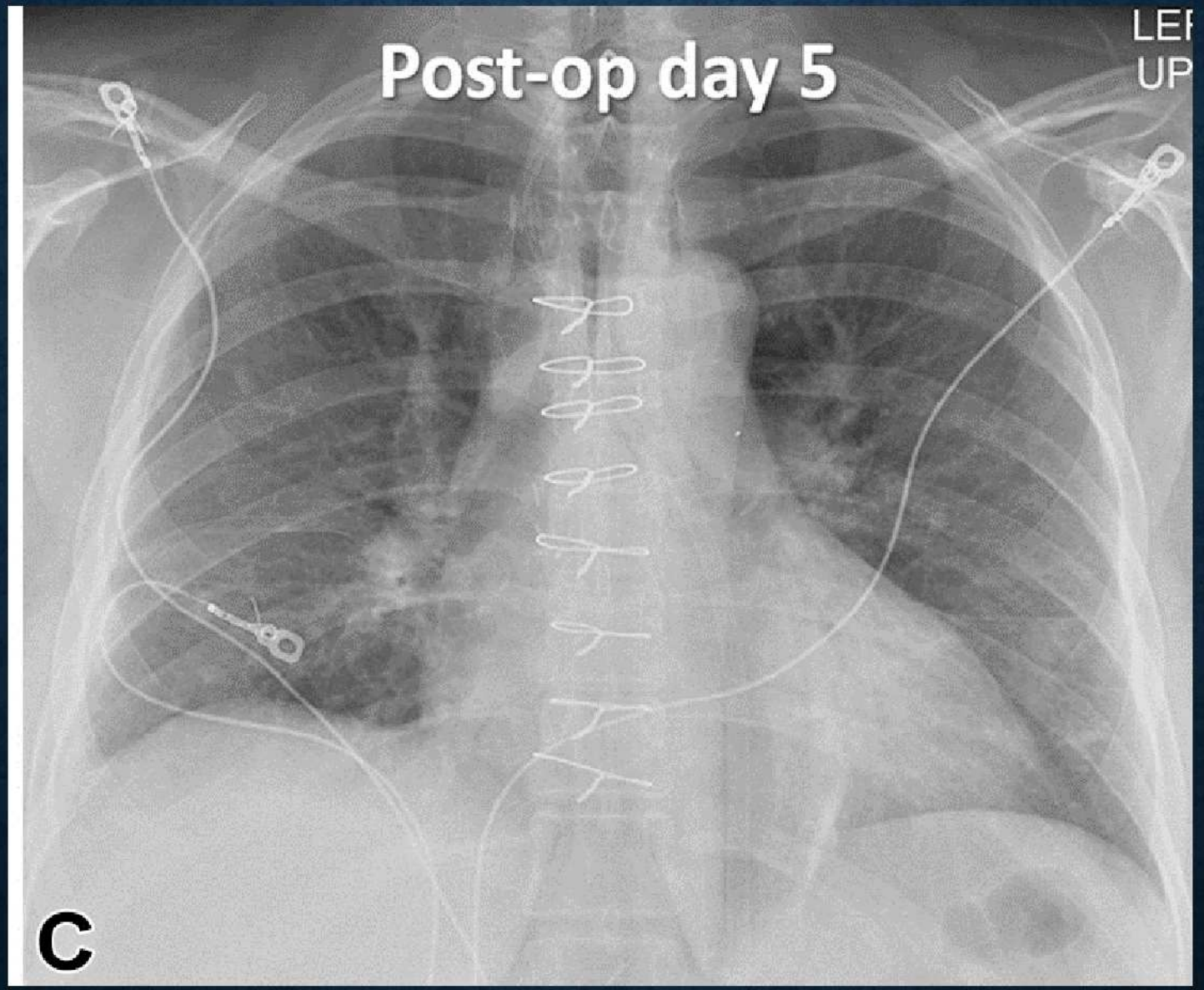

Post-op day 5
C

Table 1: Distinguishing Imaging Findings of CTEPH Mimics at CTPA

CTEPH Mimic	Features More Suggestive of a CTEPH Mimic	Features More Suggestive of CTEPH or CTEPD
Acute PE	Filling defects at center of vessels Vessel expanded Acute margin between clot and PA wall Main PA does not dilate in response to acute PE nor does RV hypertrophy	Contracted vessels, webs, and bands Obtuse margin between clot and PA wall Main PA often dilated and RV is hypertrophied in CTEPH
In situ thrombus	Peripheral thrombus in a central PA without segmental or subsegmental disease PA wall calcifications Bronchial artery collaterals uncommon in PAH Diffuse peripheral vascular pruning	Significant segmental or subsegmental thromboembolic disease, with focal areas of vascular attenuation in regions of disease
Vasculitis	Systemic arterial involvement Wall thickening Pseudoaneurysms Thin beaded vessels Upper lung predominance	Intraluminal filling defects (however, may be present within PA aneurysms [eg, in Hughes-Stovin syndrome])
Pulmonary artery sarcoma	Central filling defect with convex margin extending toward pulmonary valve or into contralateral PA (sausage sign) Expansile beaded filling defect that grows despite anticoagulation Extravascular invasion or enhancement within the intravascular component Nodal or parenchymal metastatic disease Lack of bronchial collaterals Outside of direct extension, bilateral disease uncommon	Absence of expansile clot with convex margins Bilateral disease common Bronchial collaterals Presence of chronic clot does not exclude sarcoma
Fibrosing mediastinitis	Extrinsic soft tissue compressing vessel Findings of granulomatous disease elsewhere, including nodal calcifications and granulomas Occlusions of other structures, such as ipsilateral pulmonary veins	Only intraluminal filling defects

Note.—There is overlap in imaging features, and CTEPD can coexist with its mimics. PA = pulmonary artery, PE = pulmonary embolism, RV = right ventricle.

MIDA KOJU VÕTTA

- Ärge võtke siit midagi koju
- Kui tahate, võtke hea tuju koju

OLULIST

- Multidistsiplinaarne lähenemine
- Saatekirjal ägedad vs kroonilised sümptomid
- Uuringute valik vastavalt vajadustele
  - Protokolli optimeerimine?
- Eriti oluline vaadata KT-l südant
- KT ei asenda kõike, aga ühe uuringuga saab kordades rohkem infot
- Tasuks meeles pidada „miimikuid“
- Kirurgiline ravi alati eelistatud kui riskid aktsepteeritavad

KIRJANDUS

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