

# How do you work up a patient with chronic thrombo-embolic pulmonary disease

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# Disclosures

Vikas Aggarwal: I have no relevant relationships with commercial interests to disclose.

Robert Frantz:

Consulting/Steering Committee: Janssen, Liquidia.

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# Case : 70 year old man, functional class III dyspnea

- Massive PE requiring air evacuation, lytic therapy 20 years ago
- A/C for 2 years, then stopped
- Diverticulitis, temp colostomy, multiple bowel operations 13 years ago
- TIA 12 years ago; PFO closed with Amplatzer device
- E coli sepsis with spinal involvement after prostate biopsy 10 years ago; prolonged hospital stay, needed spinal rods
- Recurrent PE 1 ½ years ago; dyspnea persisted despite A/C
- Persisting dyspnea prompts consideration of CTEPH



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# Evaluation

- History, exam, ECG, CXR, PFTS with DLCO, basic lab work including CBC, electrolytes, NTproBNP
- Consideration of special coagulation studies
- Echo with attention to RV
- Ventilation/Perfusion Lung scan with SPECT
- CT angiography
- 6 min walk
- Cardiopulmonary exercise testing
- Right heart catheterization
- Planar pulmonary angiography or DynaCT
- Multidisciplinary case review



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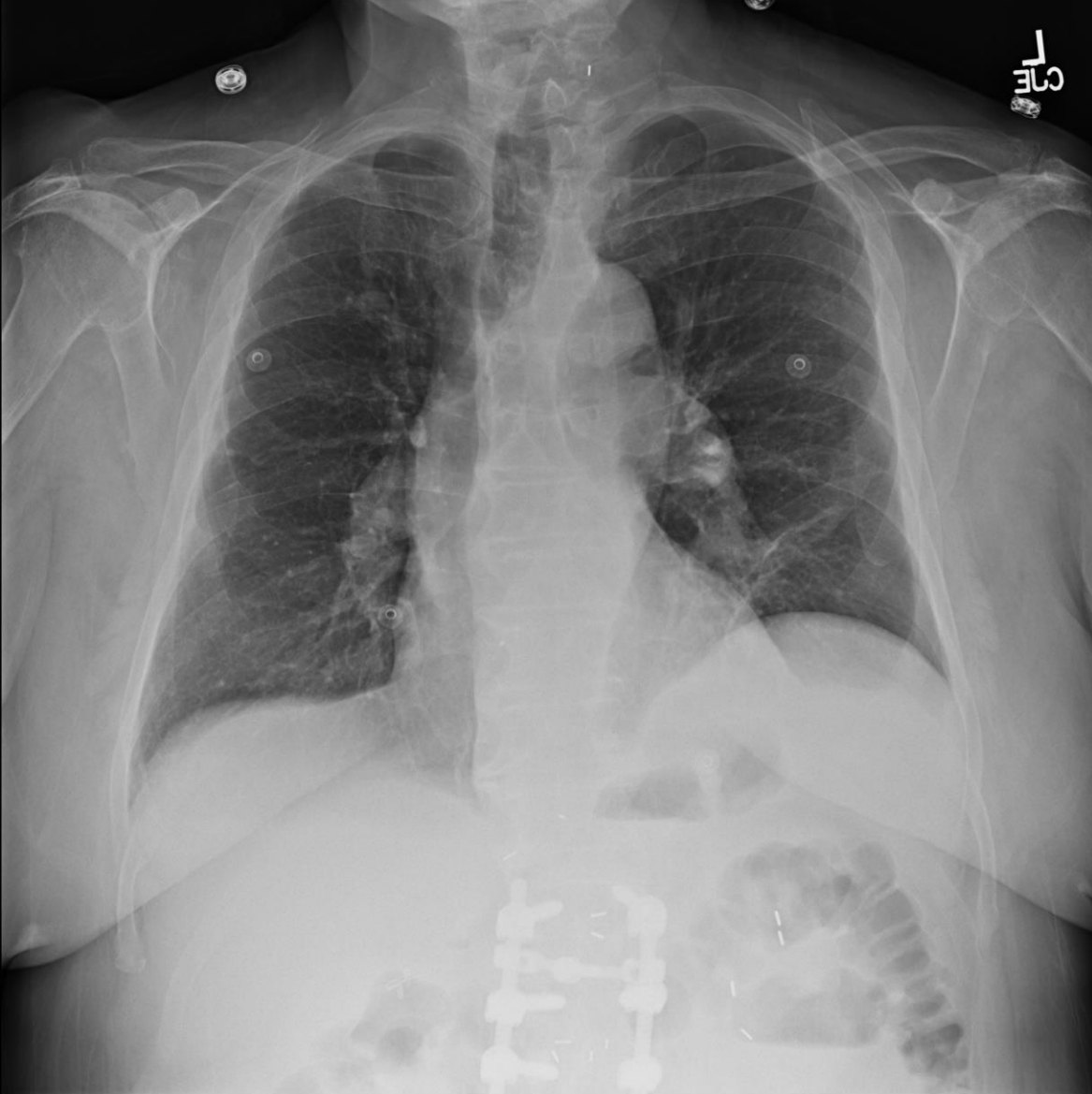
# Noninvasive Cardiopulmonary Exercise Testing

- Cardiac output response (cardiac limitation to exercise)
- Particular attention to ventilatory efficiency
  - (VE/VC02 slope)
  - With successful BPA or open surgery, ventilatory efficiency will improve due to improvement in ventilation/perfusion matching (reduced dead space ventilation)



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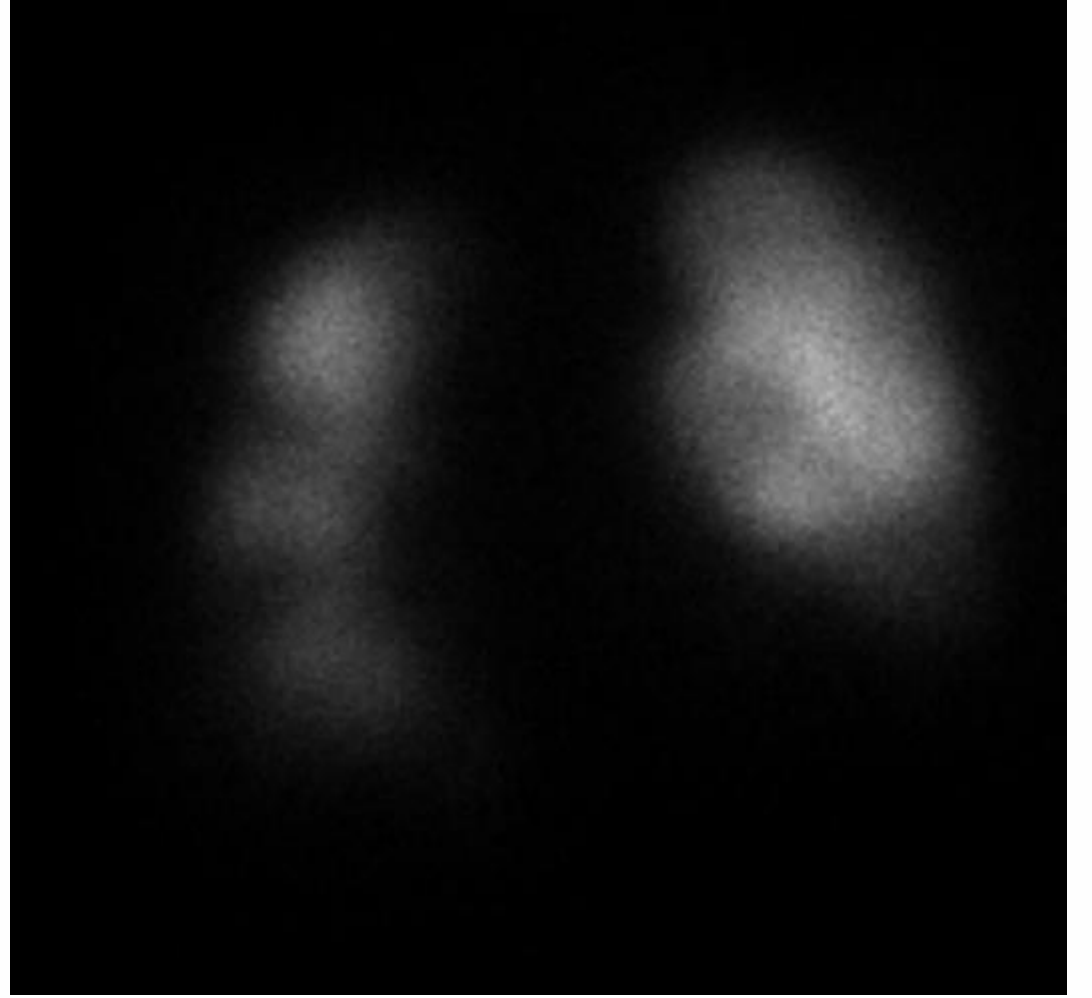
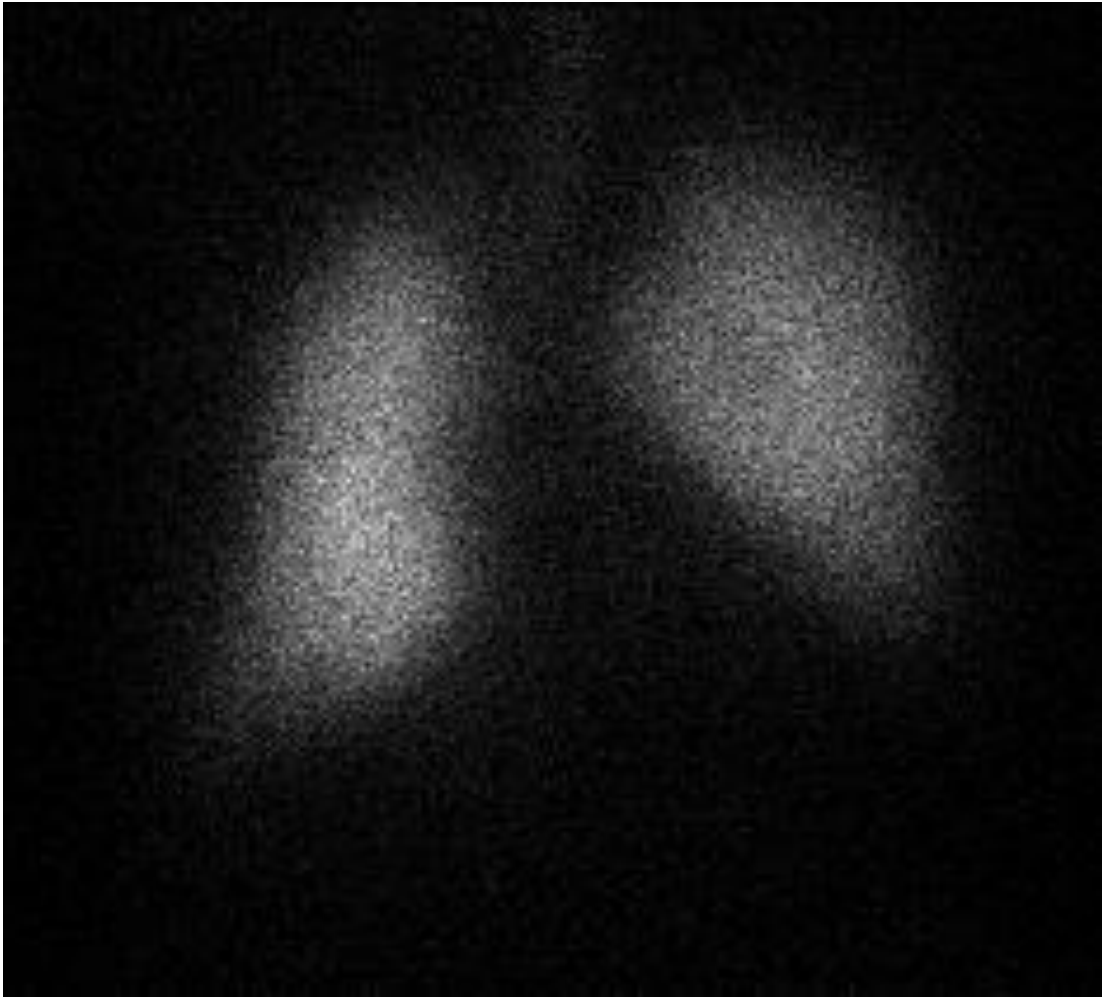
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# Ventilation/Perfusion Lung Scan

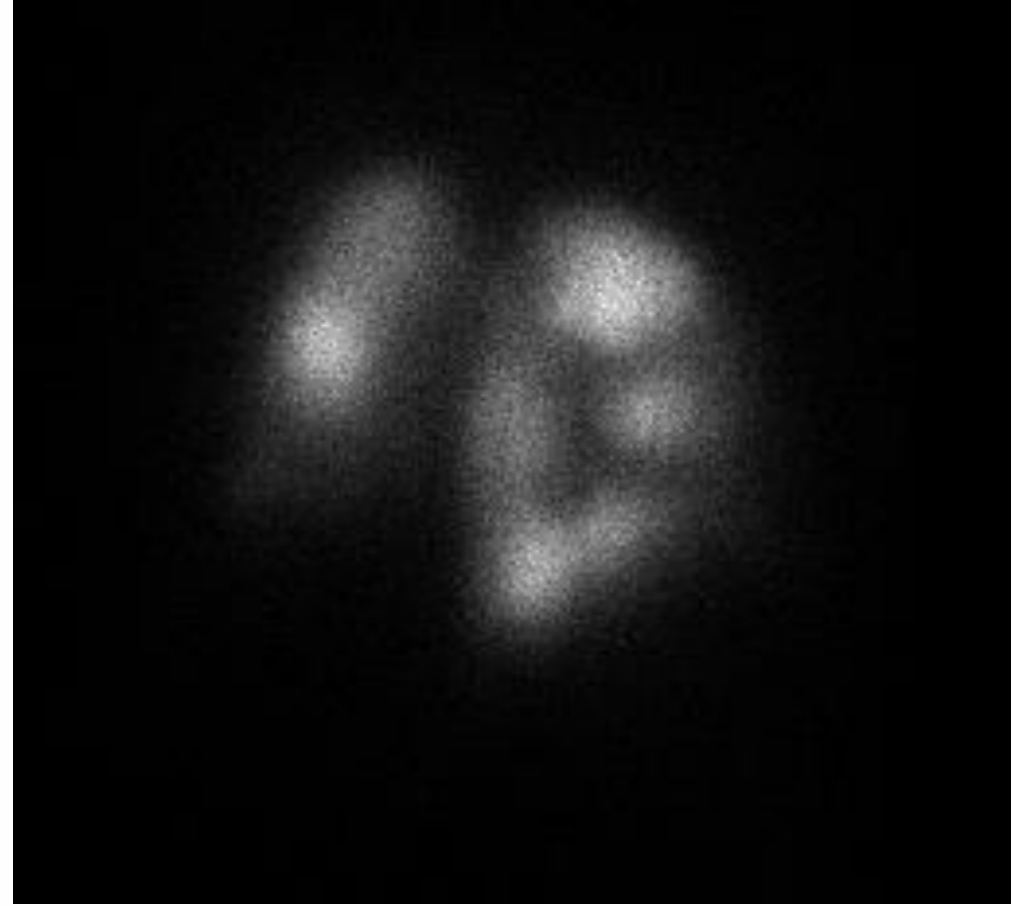
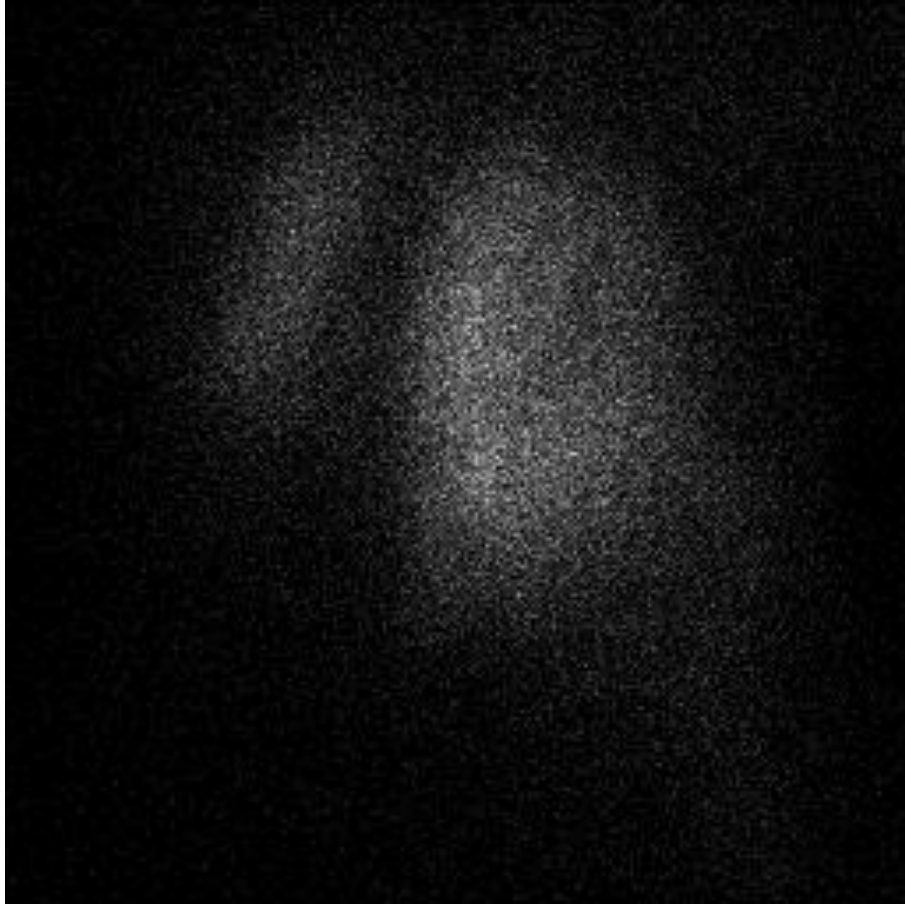


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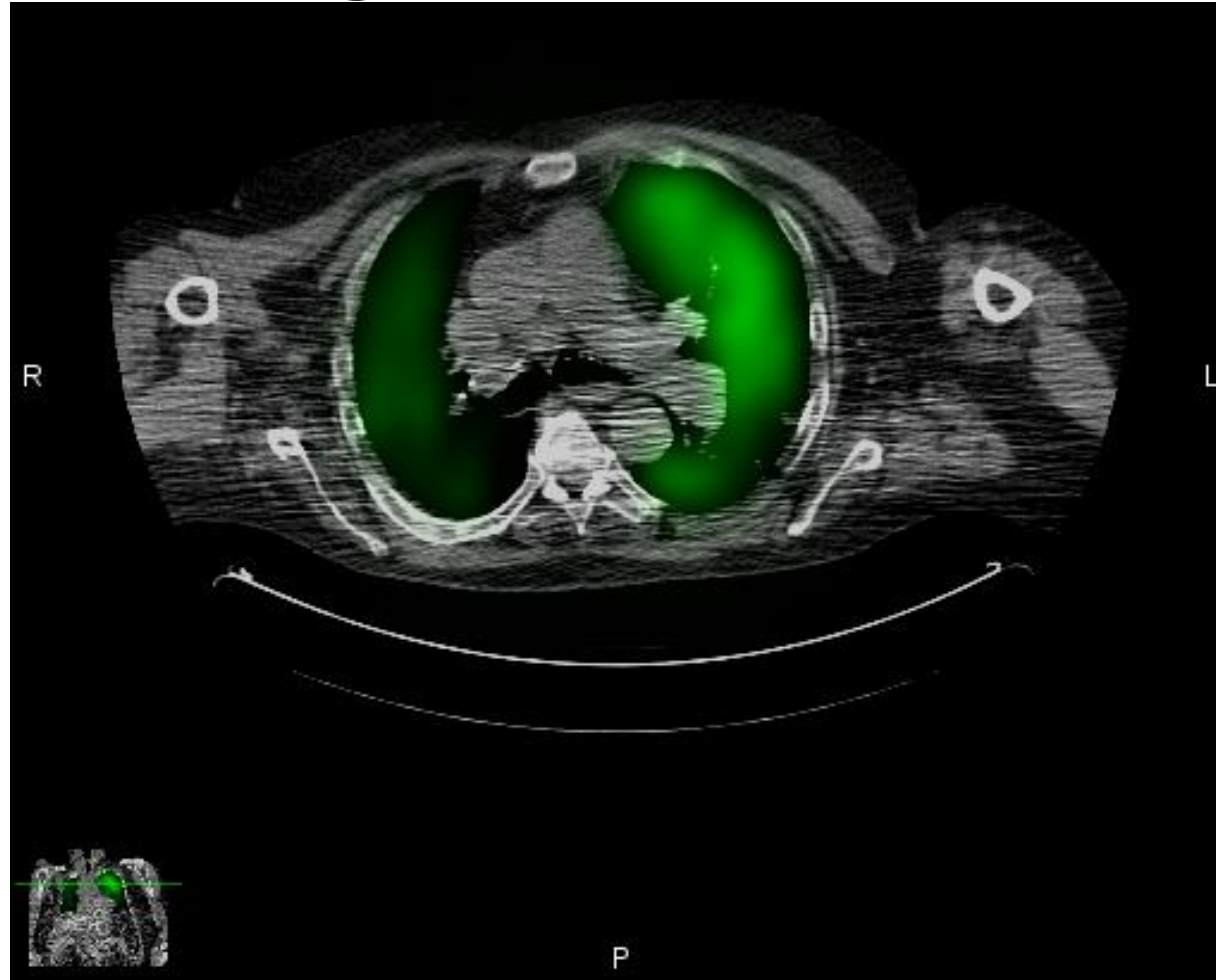
# Right Posterior Oblique V/Q Scan



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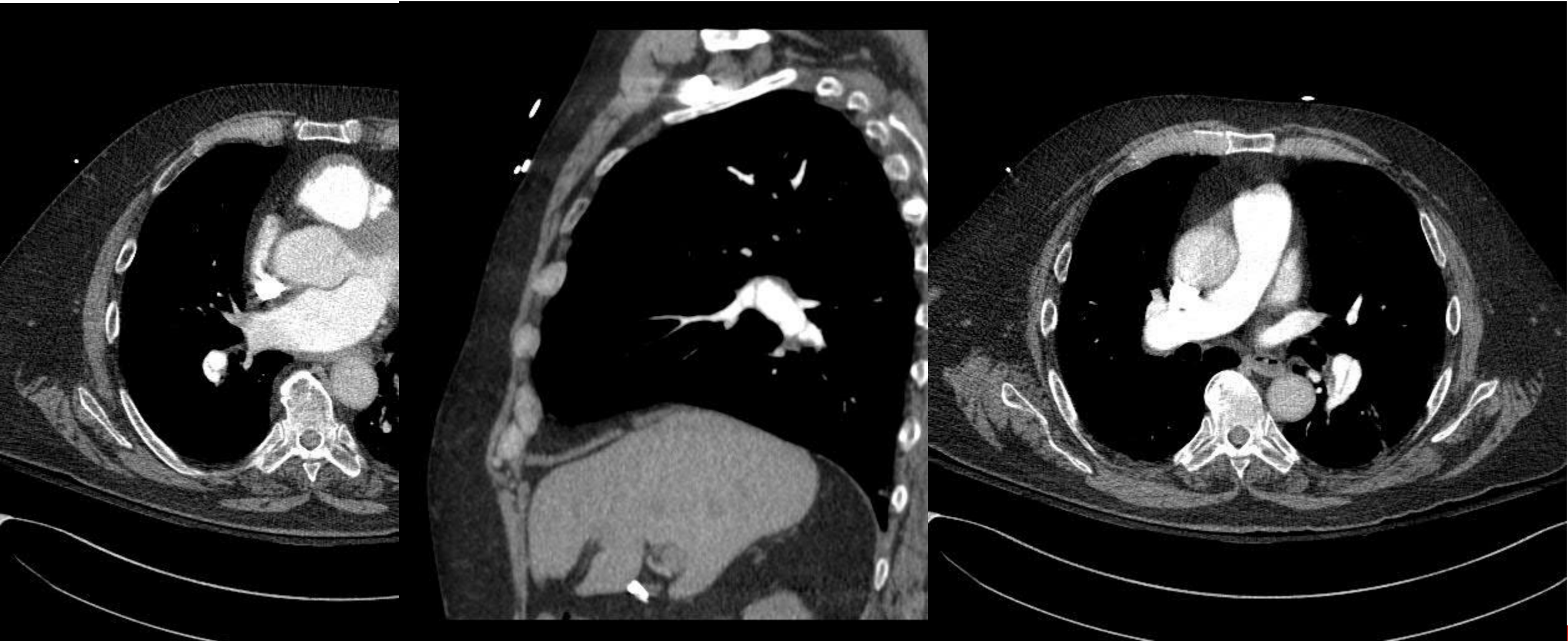
# SPECT Fusion Images



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# CT Images



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# Right Heart Catheterization

Coronary angiography if potential for open surgery

Consider iCPET if CTED, unclear relationship of symptoms and disease

- RA 8 PA 59/14/33 PCW 10 CO/CI 4.6/2.3 PVR 5 WU



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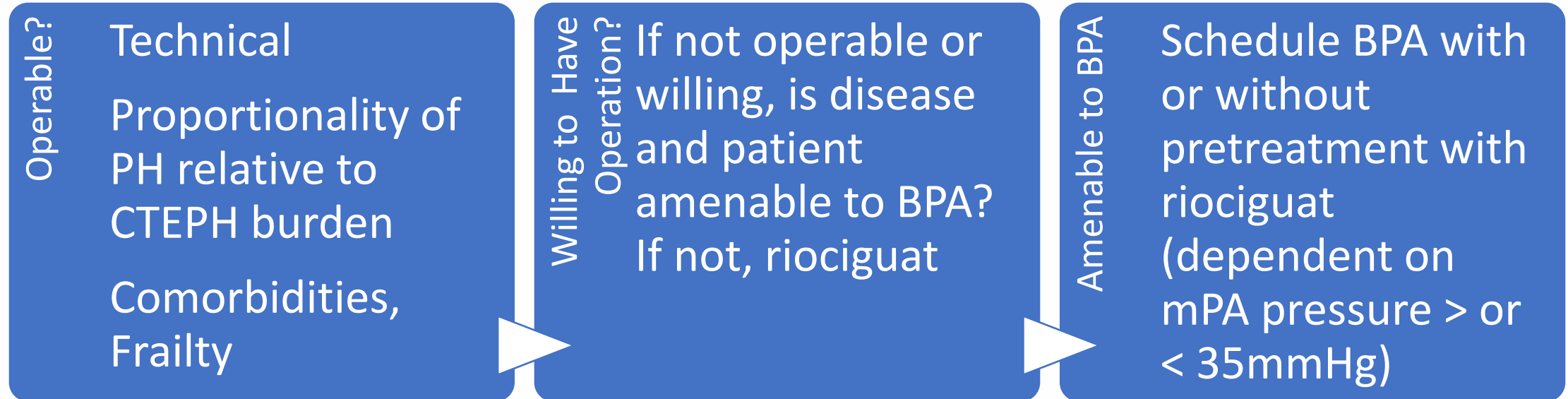
Selective  
rotational  
pulm angio  
with DynaCT



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# Pathway for Decision Making in CTEPH



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# Patients with inoperable or residual CTEPH who are considered for BPA

- If mean PA > 35, initiate riociguat before BPA
- If mean PA < 35, may proceed with BPA without use of riociguat, or may add it to assist with improvement during the BPA process



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# Our patient had two sessions of BPA separated by two months (distant travel)

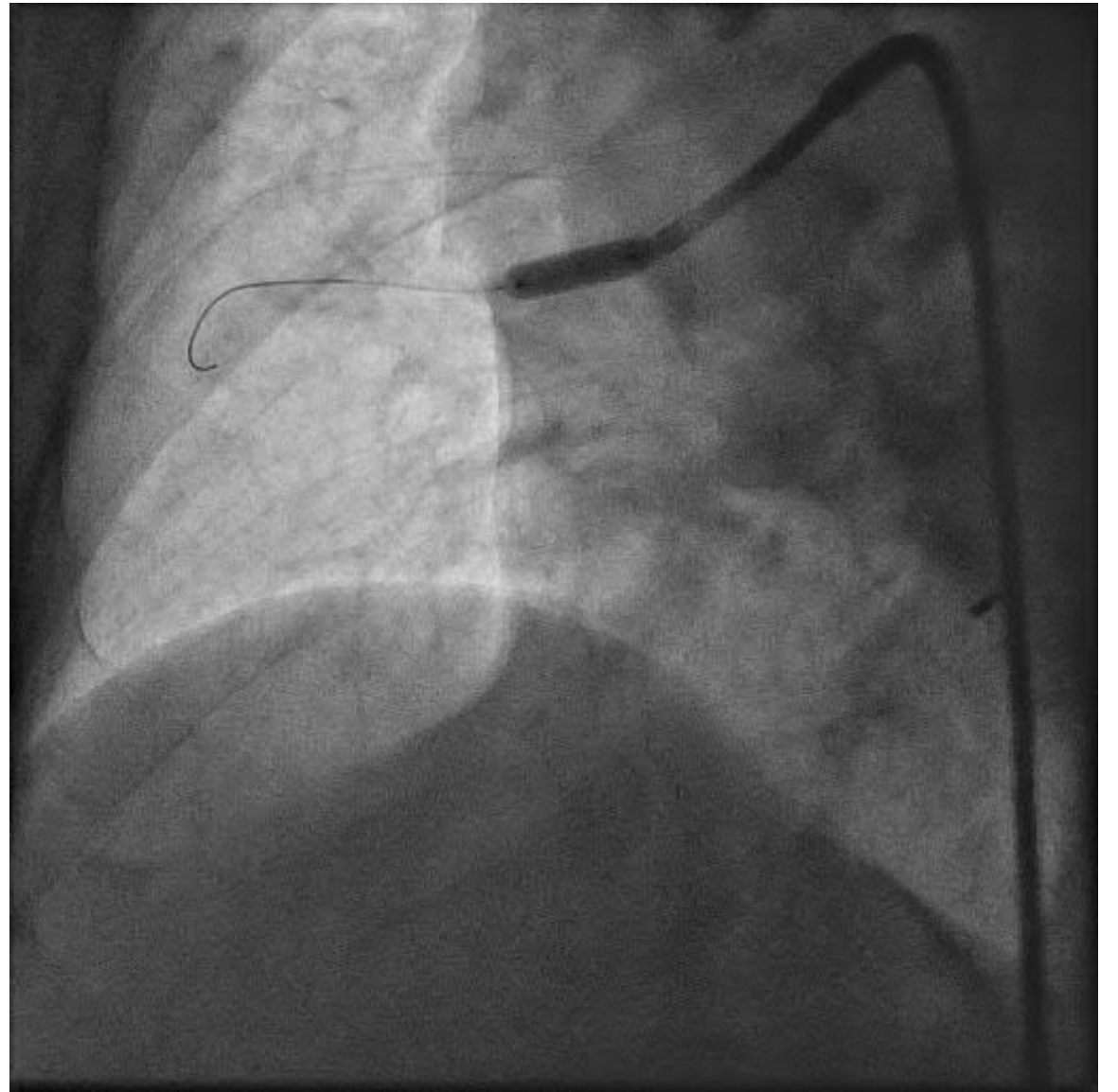
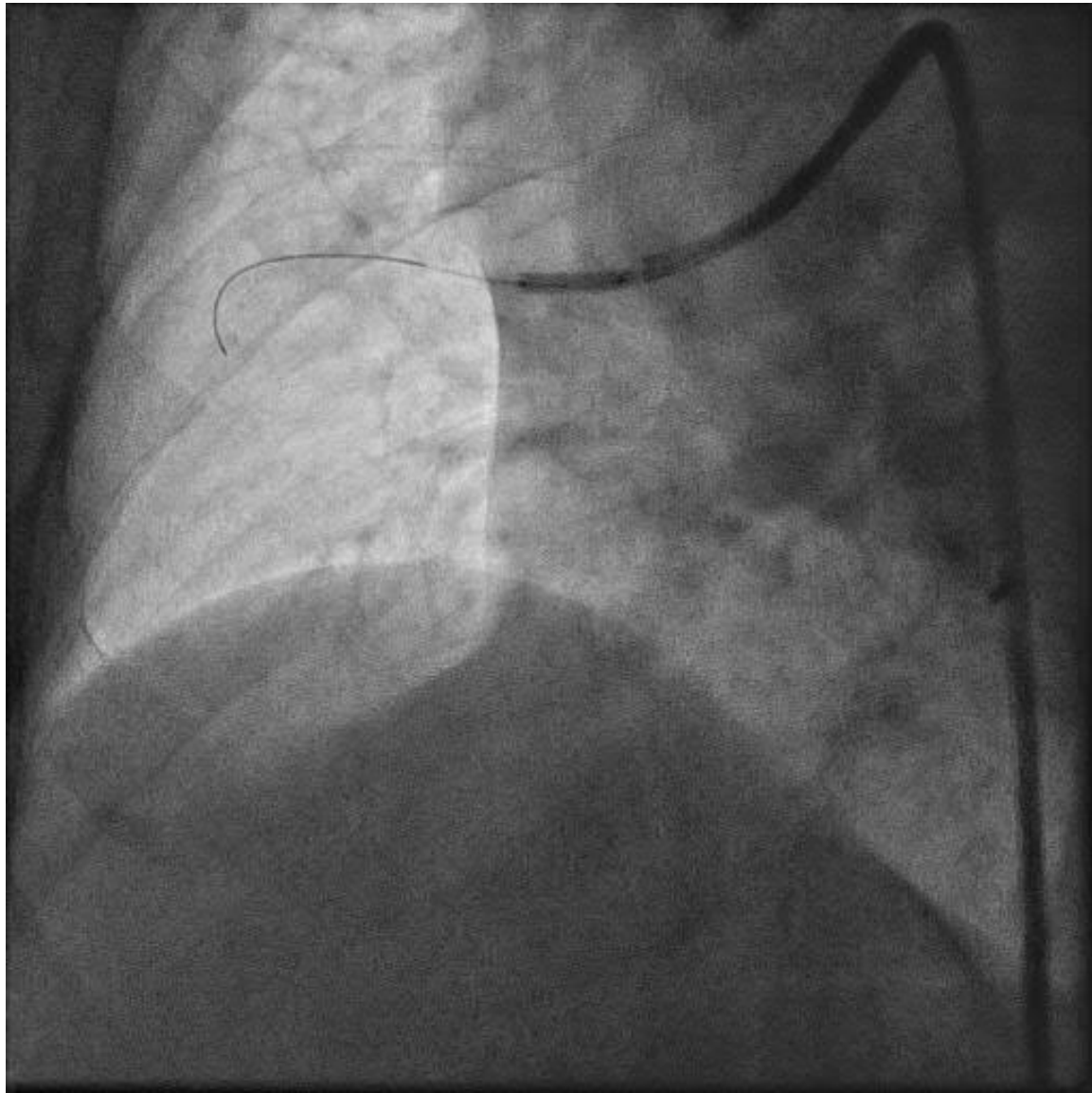
- Session 1: Right A5, A8, A9 segments
- Follow up just prior to second session:
- He has no longer been needing oxygen during the day. He has been able to be quite a bit more active in his yard, digging trenches and so forth. He will still get out of breath with walking their new dog.
- Overall he seems improved to functional class II. His VE/VCO<sub>2</sub> nadir on cardiopulmonary exercise testing today is 40, improved from 44 in July of this year. The VE/VCO<sub>2</sub> slope is 39.9, down from 41.4 in July of this year.



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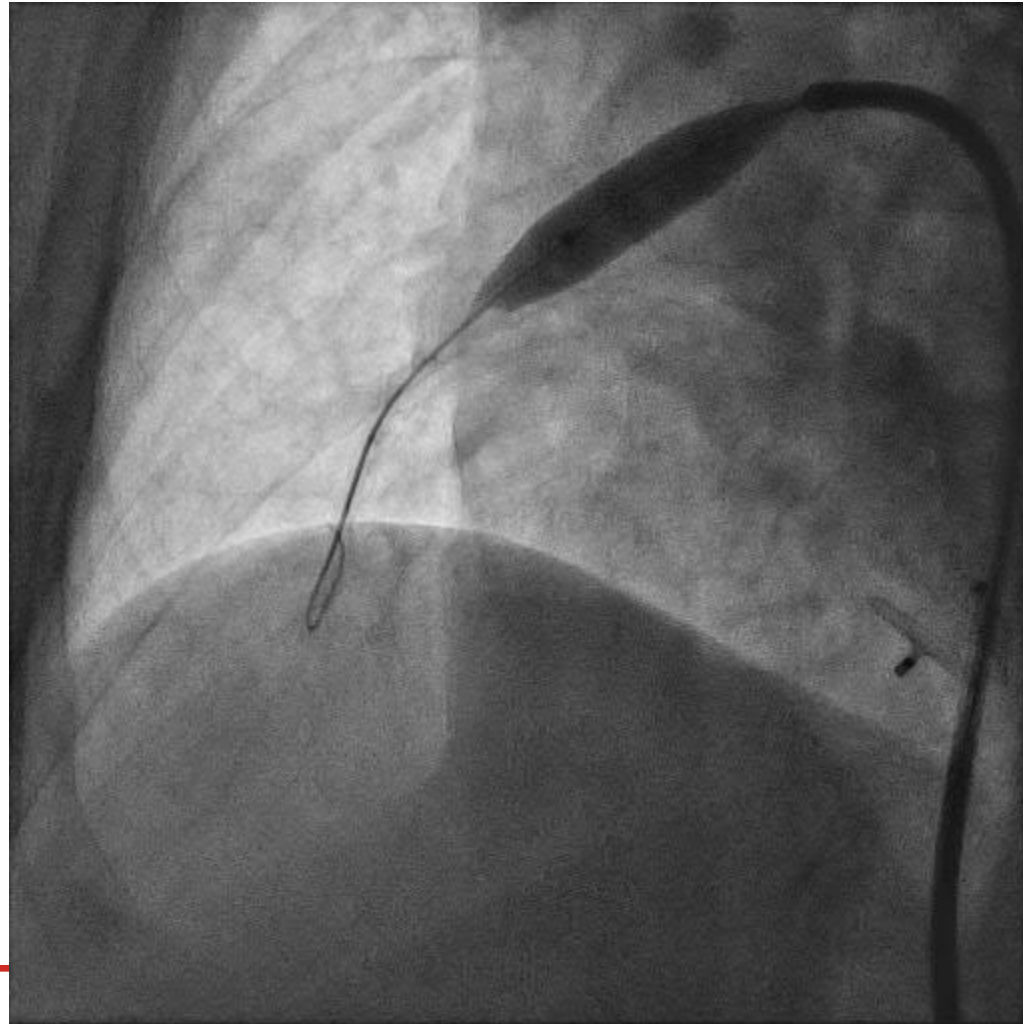


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# Balloon Pulmonary Angioplasty of Larger Segment



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# Post Procedure Care

- Hospital observation, usually overnight
- CXR am of dismissal, or sooner in event of worsening oxygenation, dyspnea, severe cough, hemoptysis
- Clear strategy for bridging anticoagulation
- Systematic reevaluation of symptoms, exercise tolerance, echo, hemodynamics, NTproBNP
- Serial sessions at least a few days apart
- Consider whether to continue or discontinue riociguat if utilized



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# Our patient had two sessions of BPA separated by two months (distant travel)

- Session 1: Right A5, A8, A9 segments
- Session 2: Right A3, A4 segments
- Felt so much better he declined to return for reassessment or additional sessions
- Continues to do well 4 years later



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# Conclusions

- Systematic evaluation of symptoms, cardiac function, exercise limitations, hemodynamics, and cardiopulmonary physiology
- Thoughtful and thorough imaging
- Multidisciplinary team-based, patient centric decision making
  
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