



Chronic Thromboembolic Pulmonary Hypertension Update 2024

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Objectives

After this presentation, the participant will be able to:

- **Identify the Risk Factors and Presentation of CTEPH**
 - Recognize the key symptoms and risk factors associated with CTEPH, including a history of pulmonary embolism and unexplained dyspnea.
- **Explain the Diagnostic Approach to CTEPH**
 - Emphasize the importance of a VQ scan as the initial test for suspected CTEPH and understand how to integrate imaging and hemodynamics into the diagnostic process.
- **Understand Multidisciplinary and Multimodal Management of CTEPH**
 - Describe the role of a multidisciplinary program in managing CTEPH, focusing on collaboration with experienced centers and the role of high-volume surgeons in optimizing outcomes.

CTEPH

- Chronic thromboembolic pulmonary hypertension
 - Complicates between 1-5% of all cases of pulmonary embolism
 - Remodeling of blood vessels after PE leading to permanent pulmonary vascular obstruction, pulmonary hypertension, heart failure, and death.
 - The only “curable” cause of pulmonary arterial hypertension
 - Standard of care is pulmonary thromboendarterectomy (PTE)
 - In the past 5-10 years the procedure balloon pulmonary angioplasty (BPA) has been optimized to provide another option for patients who are not candidates for PTE
 - There is only 1 FDA approved medical therapy – Riociguat.
 - Riociguat is only approved for patients deemed “inoperable” by a CTEPH Center

Case 1

40-Year-Old Female from the Caribbean

- **Saddle pulmonary embolism** in 2021 post-COVID-19, no thrombolysis
- Right-sided heart failure developed post-PE
- Admitted in January 2022 for bilateral leg swelling, worsening dyspnea, and pleural effusion
- **Diagnosis of CTEPH** presumptively made in 2022 with associated right-sided heart failure and abnormal CTA

Trial of Tadalafil initiated in 2022 for pulmonary hypertension

Progressively worsening dyspnea (WHO Functional Class III) by 2024

Echocardiogram (August 2024):

Moderate-large pericardial effusion



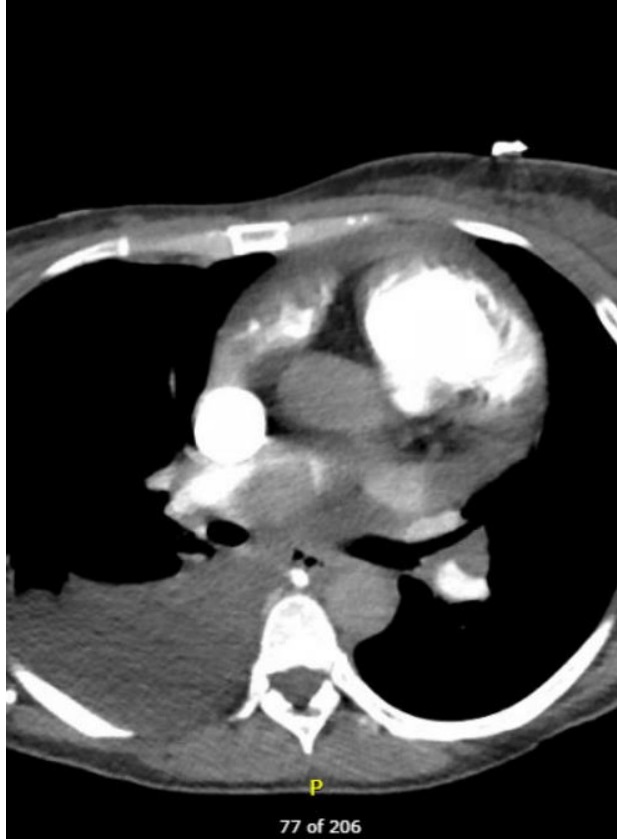
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graph TD; A[Moderate-large pericardial effusion] --> B[RSVP of 72 mmHg]; B --> C[Severely dilated RV, moderate TR, mild RV dysfunction]; C --> D[LVEF 60-65%]; D --> E[Inferior vena cava mildly dilated, large pleural effusion];
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RSVP of 72 mmHg

Severely dilated RV, moderate TR, mild RV dysfunction

LVEF 60-65%

Inferior vena cava mildly dilated, large pleural effusion



CTA Chest

- Chronic thromboembolic disease in the mid to distal right pulmonary artery
- Near-total occlusion of the basilar segments
- Severe enlargement of right-sided heart chambers



Reflux of contrast into IVC

- Eccentric thrombus in the L interlobar PA
- Chronic thromboembolic disease in the mid to distal right pulmonary artery

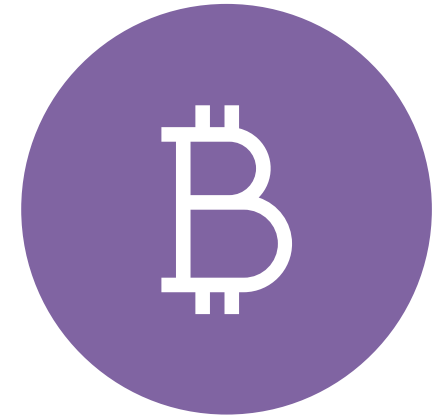
Other Findings



ATRIAL FLUTTER WITH RVR



PLEURAL EFFUSION



ELEVATED BNP (8025)

Management Plan

ADMISSION TO ICU FOR DIURESIS, TEE, CARADIOVERSION



PLAN FOR RIGHT HEART CATHETERIZATION AND PULMONARY THROMBECTOMY EVALUATION



CONTINUATION OF TADALAFIL, ANTICOAGULATION WITH ELIQUIS, AND SUBSTITUTION OF TADALAFIL WITH RIOCIQUAT POST-DISCHARGE



Case 2

Patient: Mr. P, 60-year-old male

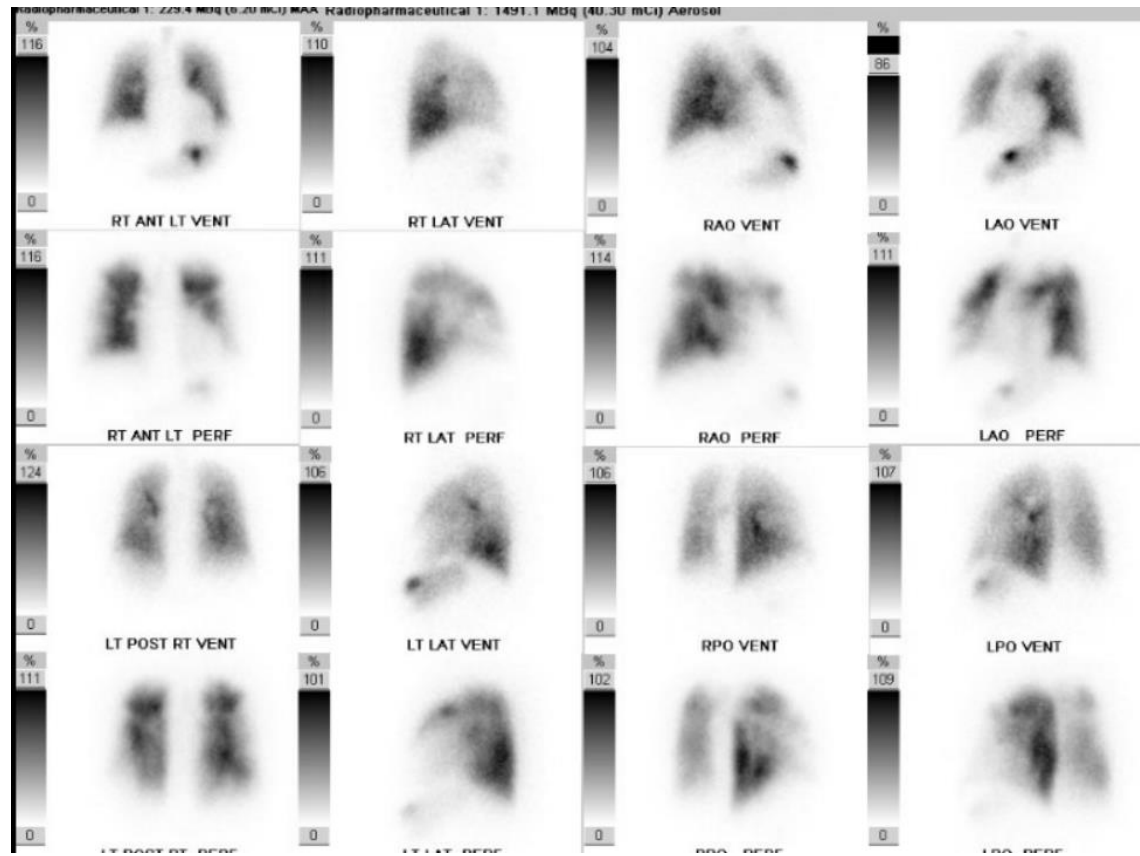
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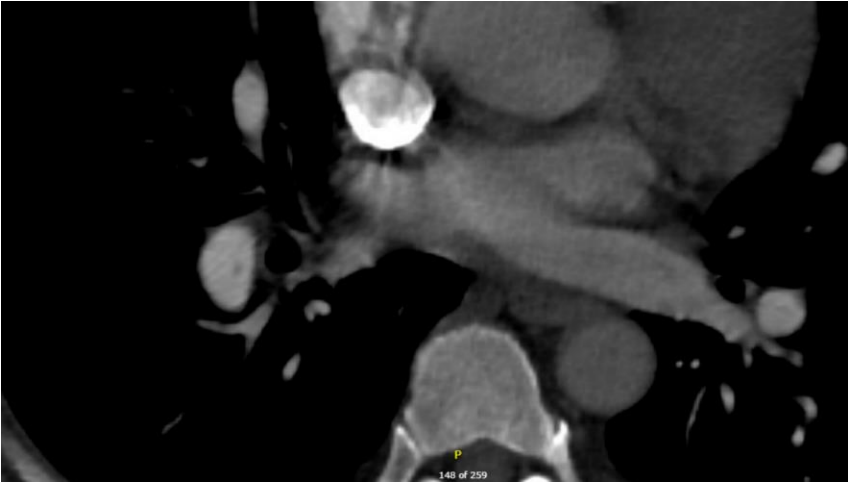
- Recurrent PE (2020, 2021) treated with lifelong anticoagulation
- Chronic hypoxemia requiring nocturnal oxygen
- Progressive shortness of breath, dizziness, and syncope

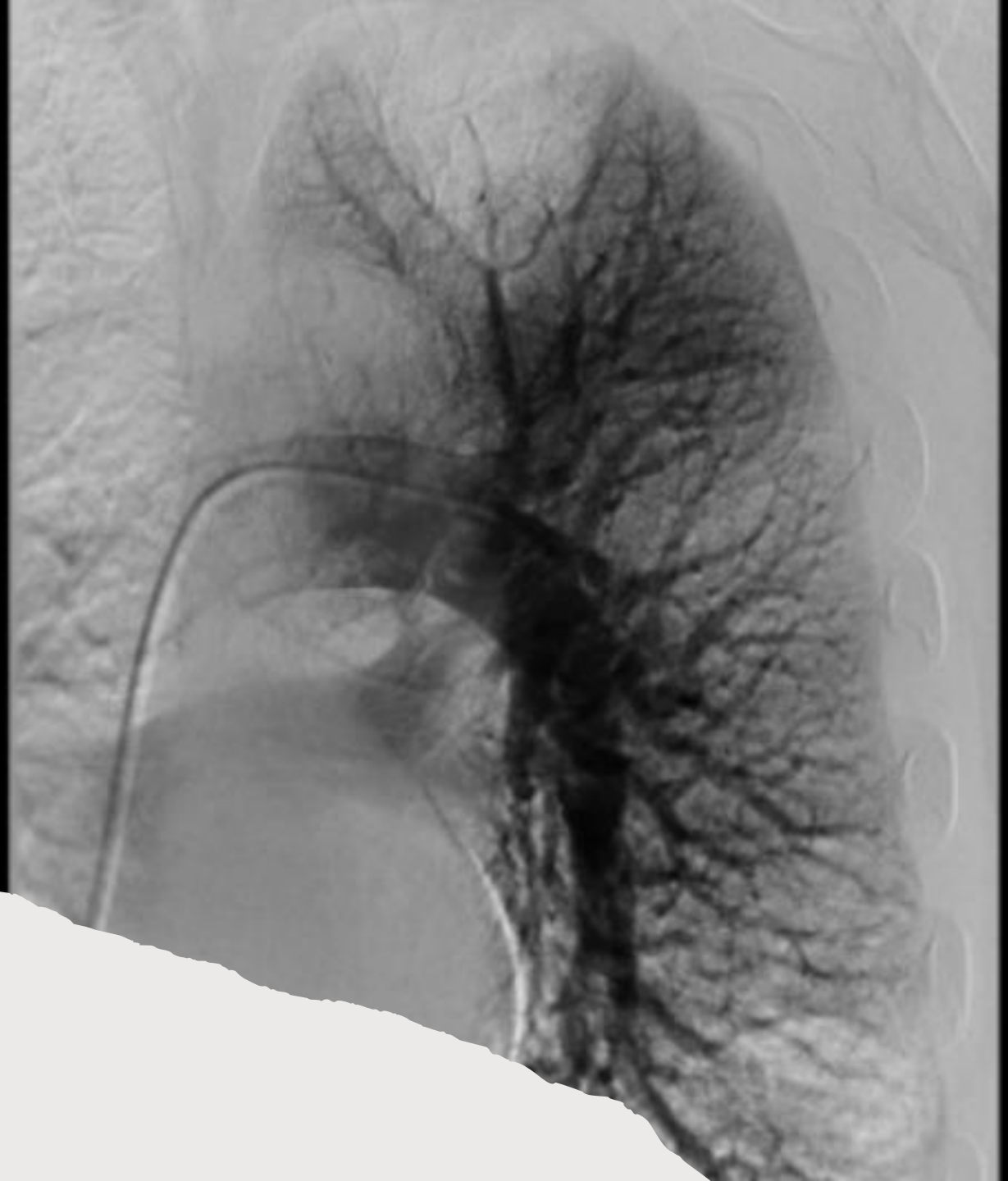
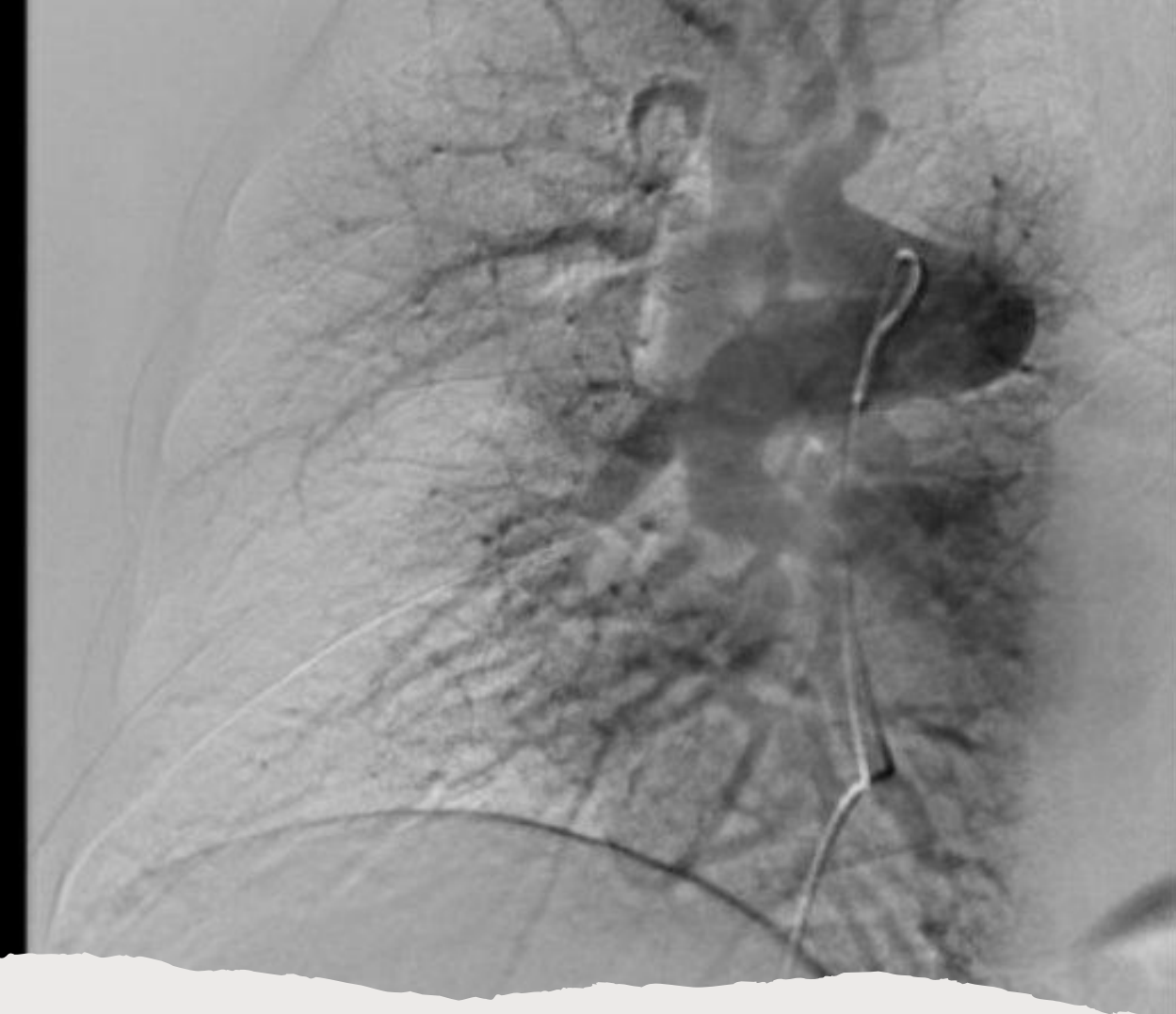
Key Investigations:

- VQ Scan (2/23): Heterogeneous perfusion, moderate-to-high probability of PE
 - Nuclear medicine suggested CTA
 - "more sensitive for PE"
- CTA Chest (2/23): No acute PE, left basilar lung nodule

VQ







Pulmonary angiogram

Significant disease with proximal involvement

Right heart catheterization

Hemodynamic Parameters	Value
Right atrial pressure	15 mmHg
Right ventricular pressure	80/22 mmHg
Pulmonary artery pressure	83/40, mean of 54 mmHg
Pulmonary capillary wedge pressure	10 mmHg
Cardiac output (Fick)	3.71 L/min

Multidisciplinary CTEPH Meeting



Date: May 25, 2023



Discussion:



- Reviewed PAgram, echo, right heart cath, and imaging
- Right upper lobe disease and mid to distal disease on the left



Decision: Proceed with PTE after consultation with Drs. Tong and Heresi. Left heart catheterization needed.

Surgery and Results

Date: June 9, 2023



Procedures:

- PTE: 4g left, 5g right
- TVr: 34 Contour band



Outcomes:

- Postop LVEF: Normal
- RVF: Normal but dilated
- Started on Eliquis POD #3

Endarterectomy
Specimen



Post-Operative Course

- Developed hypoxemia post-op due to steal syndrome, gradually improved
- Significant reduction in dyspnea; able to walk 10-15 minutes on level ground
- Mentally clearer, back to near baseline

Parameter	Value
PAS/PAD (PAM)	36/13 (21) mmHg
Central venous pressure (CVP)	6 mmHg
Cardiac output (Fick)	5.0 L/min
Cardiac index (Fick)	3.2 L/min/m ²
Systemic vascular resistance (SVR)	892 dynes·sec/cm ⁵

Recurrence and Multimodal Therapy

Recurrence:

- Patient presents with new symptoms of dyspnea on exertion.

Repeat hemodynamic studies:

- Pulmonary hypertension identified once again.
- **Presented again to CTEPH Team**

Referred for balloon pulmonary angioplasty (BPA).

Balloon Pulmonary Angioplasty (BPA)

Procedure Date: August 26, 2024

Procedure Details:

Successful cannulation of the interlobar artery branch of the left lower lobe

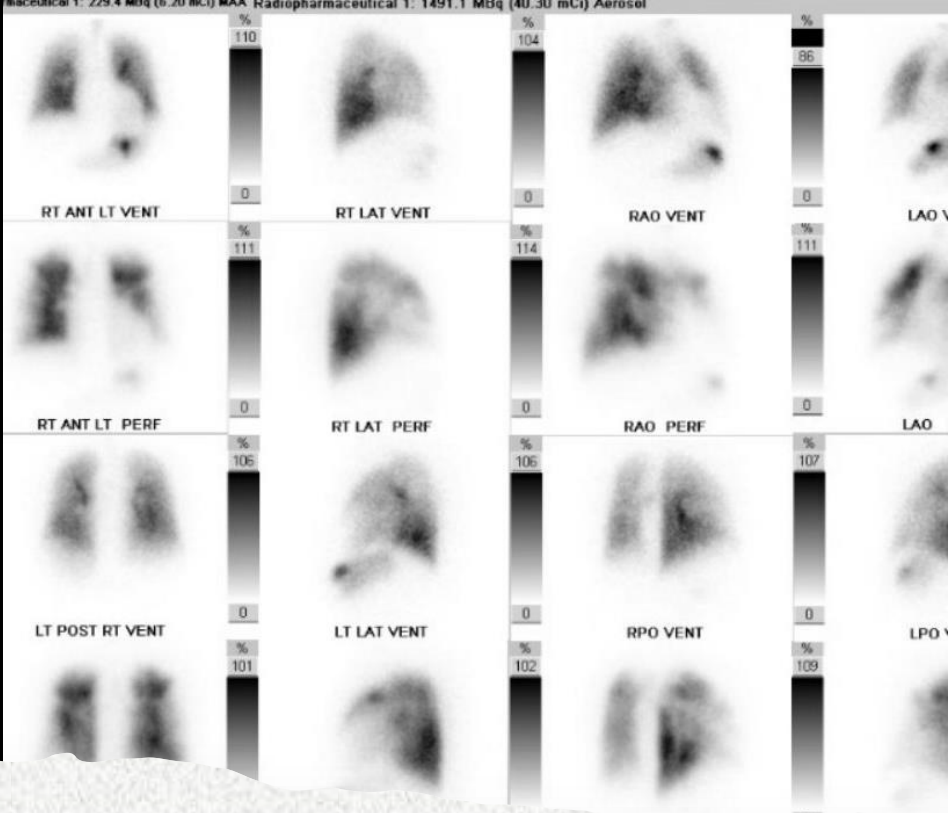
- **Angiogram Findings (Pre-BPA):**
- Significant narrowing in the left A8 and A7 branches of the lower lobe, with delayed antegrade flow and reduced perfusion.
- **Angioplasty:**
- Angioplasty performed on the anterior basal segment (A8) and medial basal segment (A7) with improved antegrade flow.
- **Angiogram Findings (Post-BPA):**
- Marked improvement in perfusion and flow in the left A8 and A7 branches, with no complications.

Hemodynamics:

Parameter	Pre-BPA	Post-BPA
Pulmonary artery pressure	43/12 mmHg	36/13 mmHg
Mean pulmonary artery pressure	23 mmHg	21 mmHg



PA pressure May 2024
45/16 (27)



PA pressure August 2024
25/7 (16)

Initial
83/40 (54) mmHg

Multimodal Therapy

- This patient received a combination of **PTE**, **BPA**, and medical therapy with **Riociguat**
 - Pulmonary thromboendarterectomy (PTE)
 - Balloon pulmonary angioplasty (BPA)
 - Medical therapy with **Riociguat**

This highlights the evolving approach in treating CTEPH, aiming for a comprehensive treatment plan that includes surgery, angioplasty, and medical management.

CTEPH Guidelines

- PTE should be offered to all eligible patients with CTEPH. Most patients experience significant improvements in symptoms and hemodynamics.
- The international registry of incident cases of CTEPH reported a 3-year survival of 90% in those receiving PTE versus *70% in those who did not have surgery*.
- Patients who have operable CTEPH should be referred for surgery without delay
- 63% of patients with CTEPH are candidates for PTE

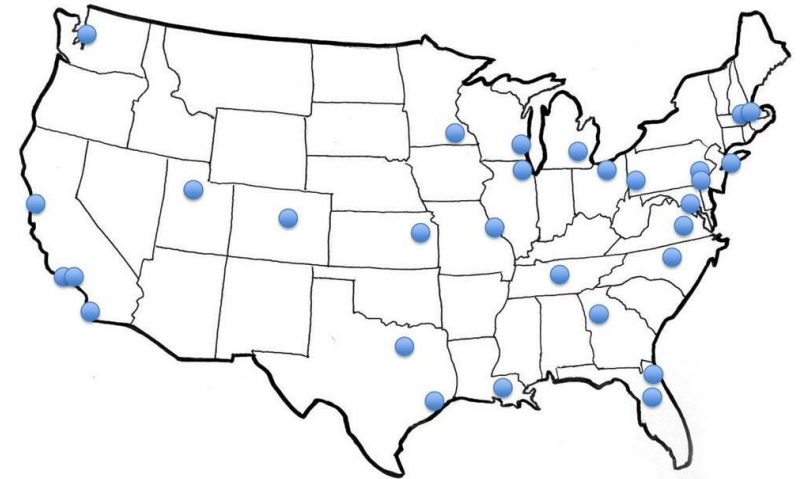
Pulmonary thromboendarterectomy

- PTE surgery is a complex bilateral procedure, which requires median sternotomy, cardiopulmonary bypass, deep hypothermia, and periods of circulatory arrest
- PTE allows for the removal of obstructing lesions, resulting in an improvement and often normalization of pulmonary hemodynamics
- Rates of residual pulmonary hypertension (PH) following PTE have been reported at 10% to 35%



CTEPH Centers

- 28 Centers are part of the national CTEPH Registry
- Only 10-15 of these sites actually offer PTE – the rest of the sites have relationships with surgical centers to refer patients
 - Emory University, LSU, Aurora, etc
- Other sites have been only recently begun PTE programs
 - Uchicago, UPMC,



CTEPH Care

- Multidisciplinary
 - Cardiothoracic surgery
 - PTE surgery, ECMO
 - Cardiac Anesthesia
 - Complex anesthetic and post-operative care
 - 20 minute intervals of ***complete hypothermic circulatory arrest***
 - Critical Care
 - Pulmonology
 - PAH expertise, drug management, cardiopulmonary exercise testing
 - Cardiology
 - Hemodynamic evaluation, cardiac imaging
 - BPA
 - Radiology
 - Nuclear Medicine (VQ/SPECT)
 - IR – Pulmonary Angiography, BPA
- Hematology
 - Evaluation of coagulation disorders
- Comprehensive rehabilitation
 - Post-surgical rehabilitation an essential part of long-term outcomes

Learning curve - Outcomes

- Multiple studies demonstrate a distinct learning curve
- Mortality overall (global) 5.6% but once the learning curve was overcome, this dropped to 2.6%.
- Some studies show that the learning curve was felt to encompass the first forty-six cases.
- However, in a study of almost 500 patients, Miyahara *et al.* noted decreasing mortality with each 100-patient block.
- Learning curve likely not only affects surgeons taking on a new, challenging procedure but the whole multidisciplinary team including anesthetic, perfusion, intensive care and ward staff



Questions?