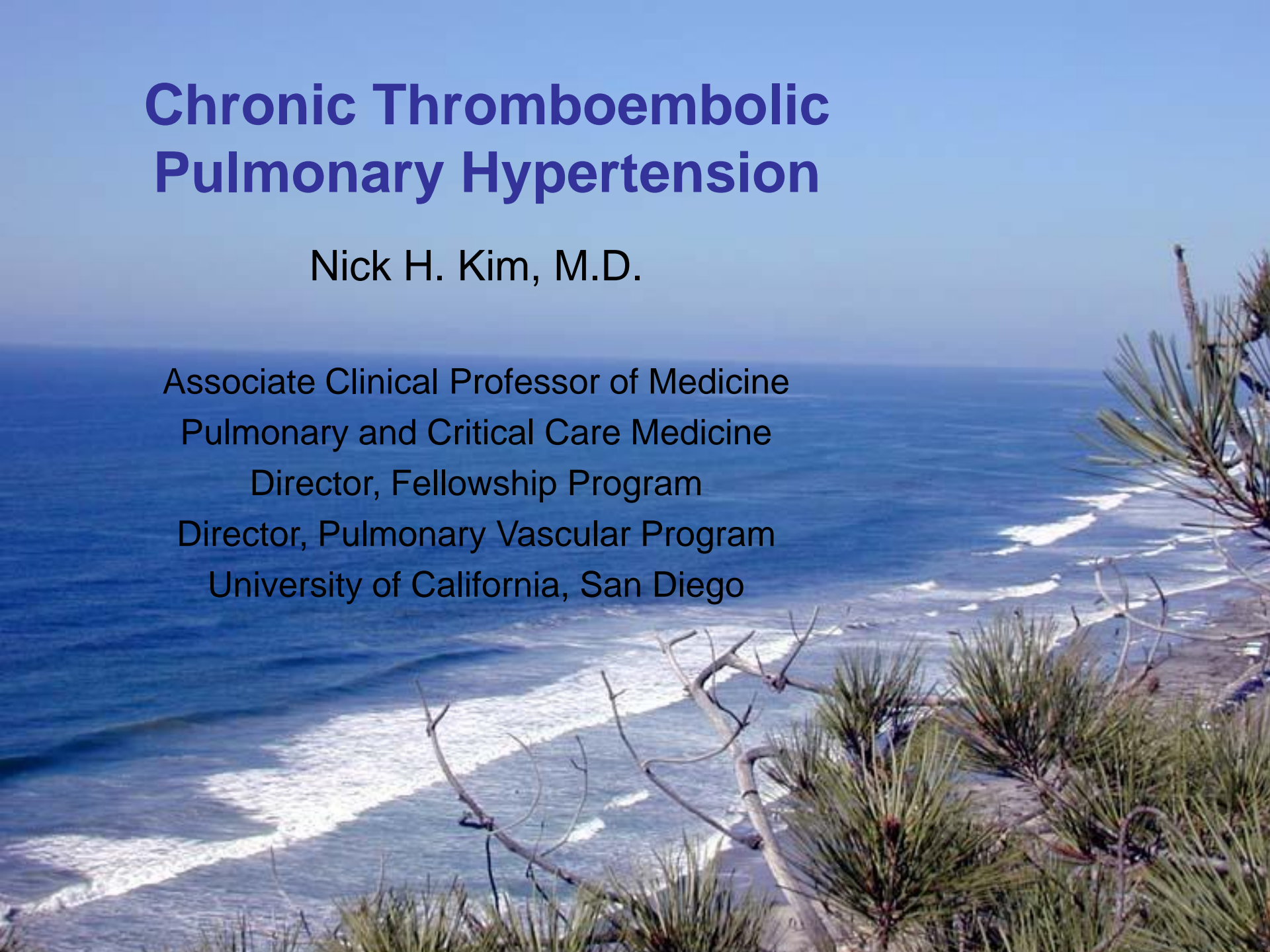


Chronic Thromboembolic Pulmonary Hypertension

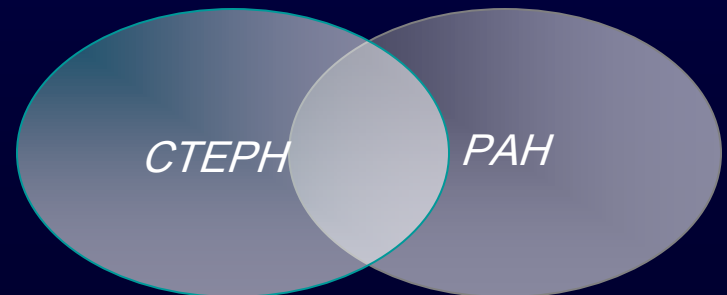
Nick H. Kim, M.D.

Associate Clinical Professor of Medicine
Pulmonary and Critical Care Medicine
Director, Fellowship Program
Director, Pulmonary Vascular Program
University of California, San Diego

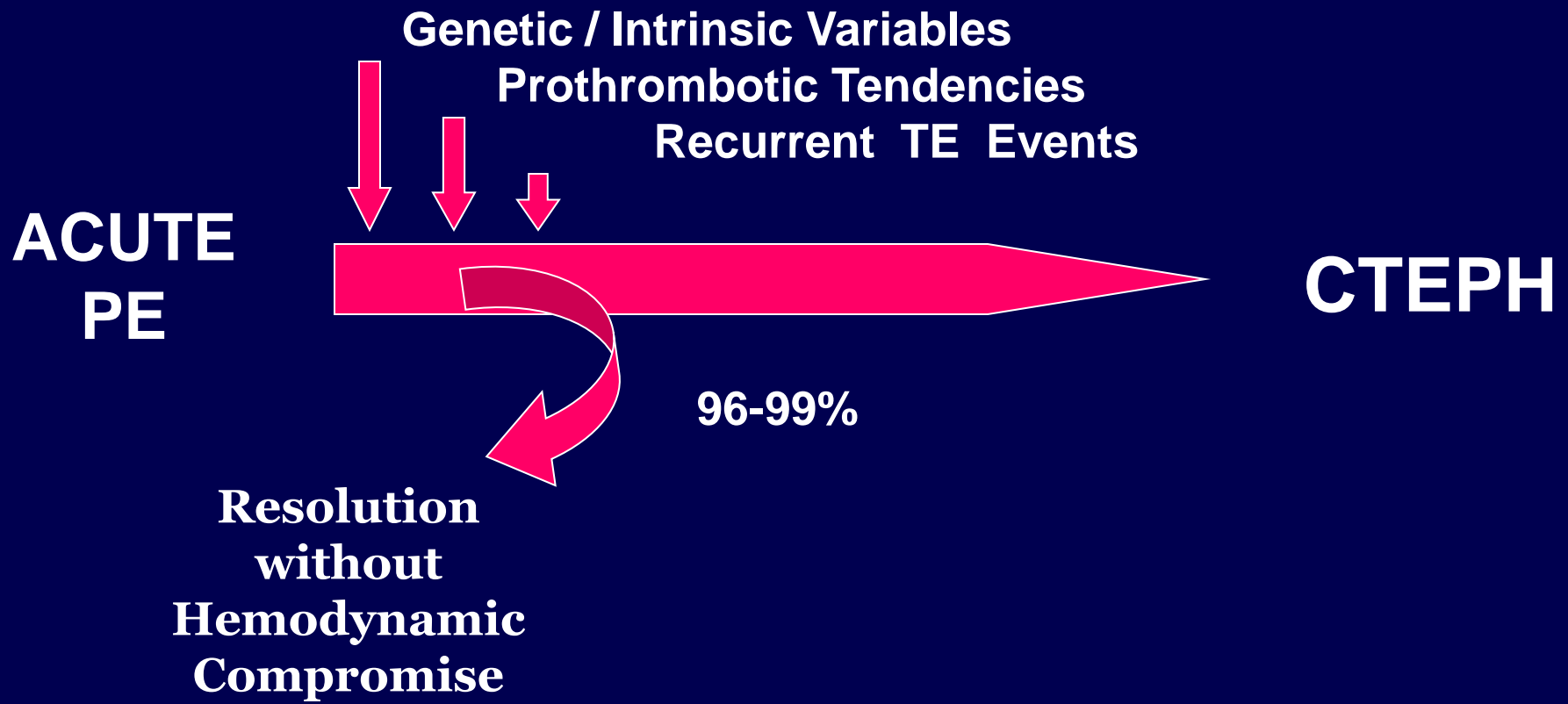


Disclosures

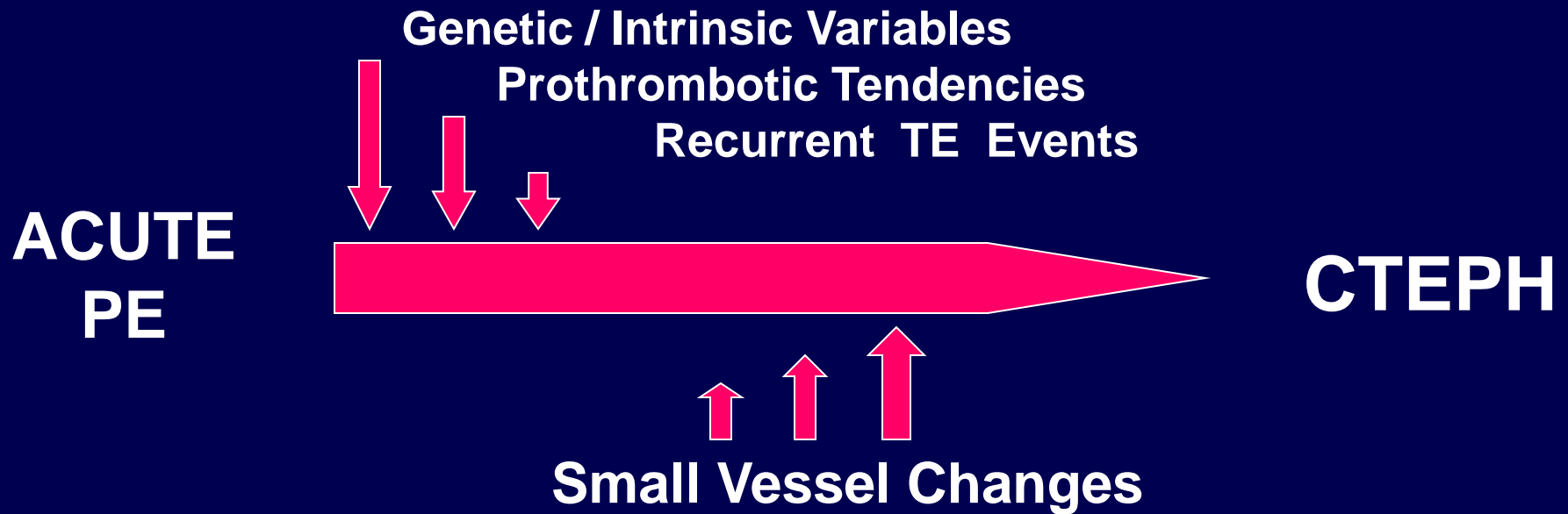
- **Research Support:**
Actelion, Gilead, United Therapeutics
- **Consultancy:**
Bayer



Natural History of Chronic Thromboembolic Pulmonary Hypertension



Natural History of Chronic Thromboembolic Pulmonary Hypertension



ssion - not intended for diagnosis

6

GROUP



ot intended for diagnosis



80BPM

14CM
68HZ

P T R
1.6 3.2

Clinical Classification of Pulmonary Hypertension (Dana Point 2008)

1. PAH

- Idiopathic PAH
- Heritable
- Drug- and toxin-induced
- Persistent PH of newborn
- Associated with:
 - CTD
 - HIV infection
 - portal hypertension
 - CHD
 - schistosomiasis
 - chronic hemolytic anemia

1' . PVOD and PCH

2. PH Due to Left Heart Disease

- Systolic dysfunction
- Diastolic dysfunction
- Valvular disease

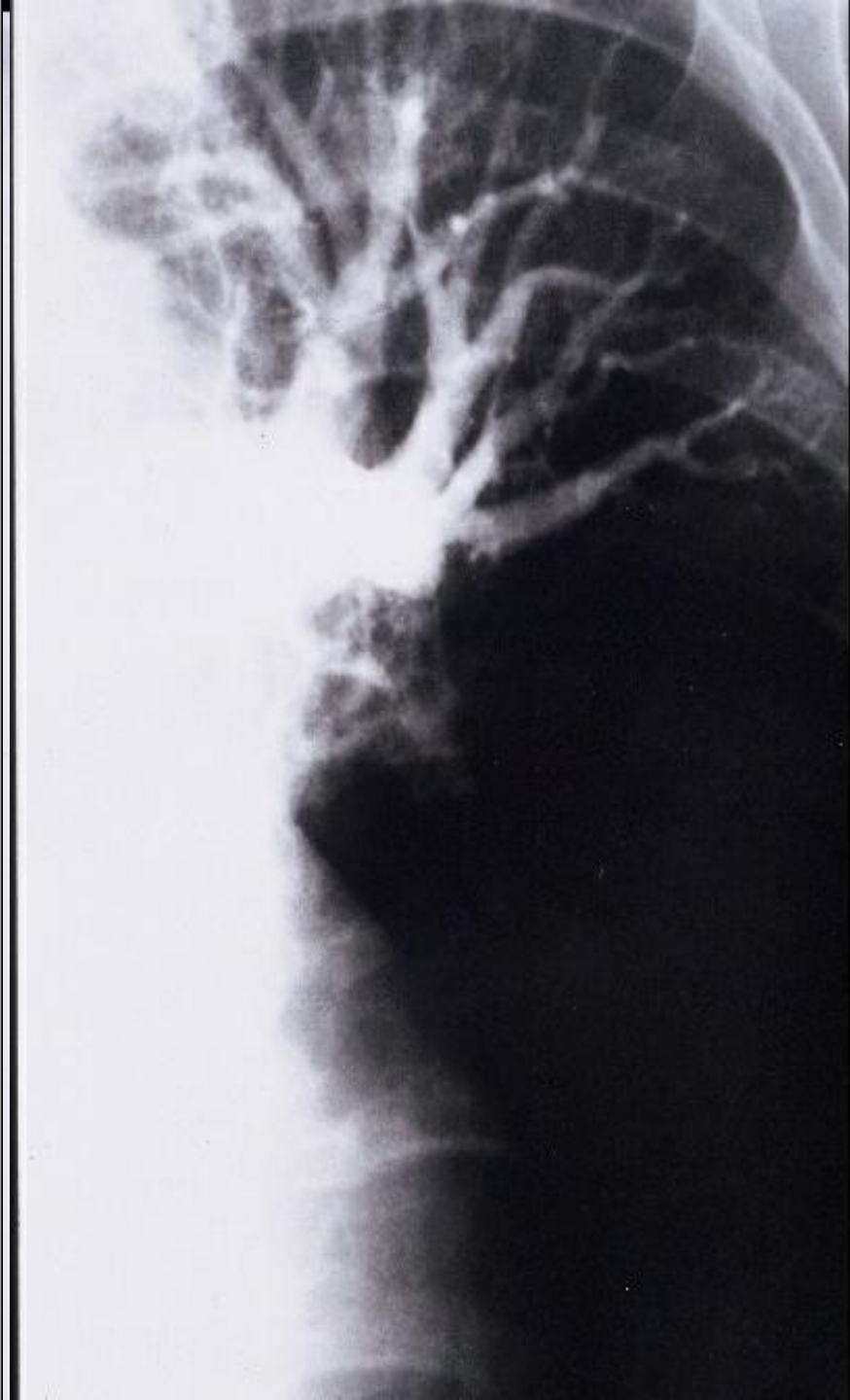
3. PH Due to Lung Diseases and / or Hypoxia

- COPD
- ILD
- Other pulmonary diseases with mixed restrictive and obstructive pattern
- Sleep-disordered breathing
- Alveolar hypoventilation disorders
- Chronic exposure to high altitude
- Developmental abnormalities

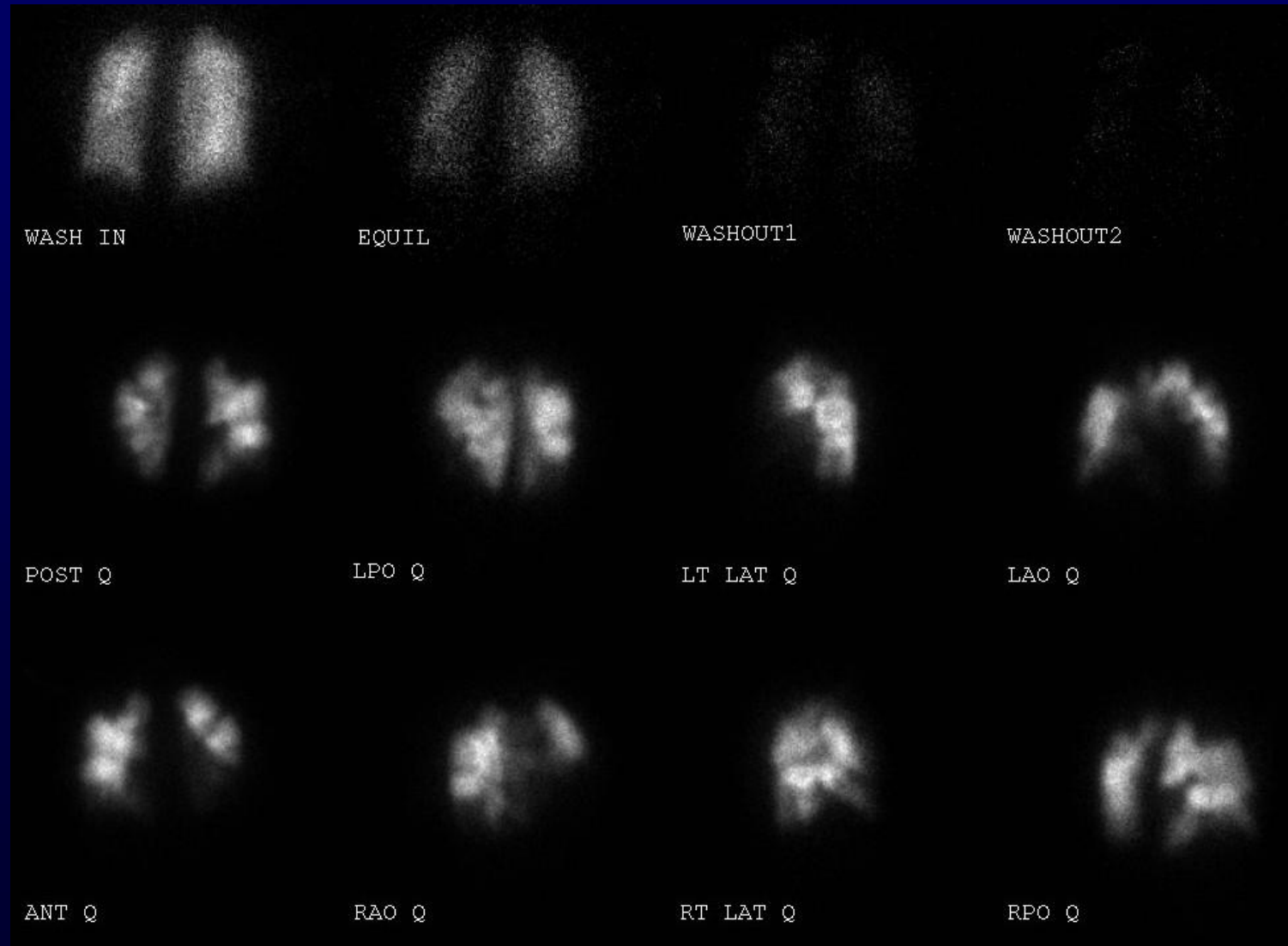
4. CTEPH

5. PH With Unclear Multifactorial Mechanisms

- Hematologic disorders
- Systemic disorders
- Metabolic disorders
- Others



VQ Scan: Screening Test of Choice



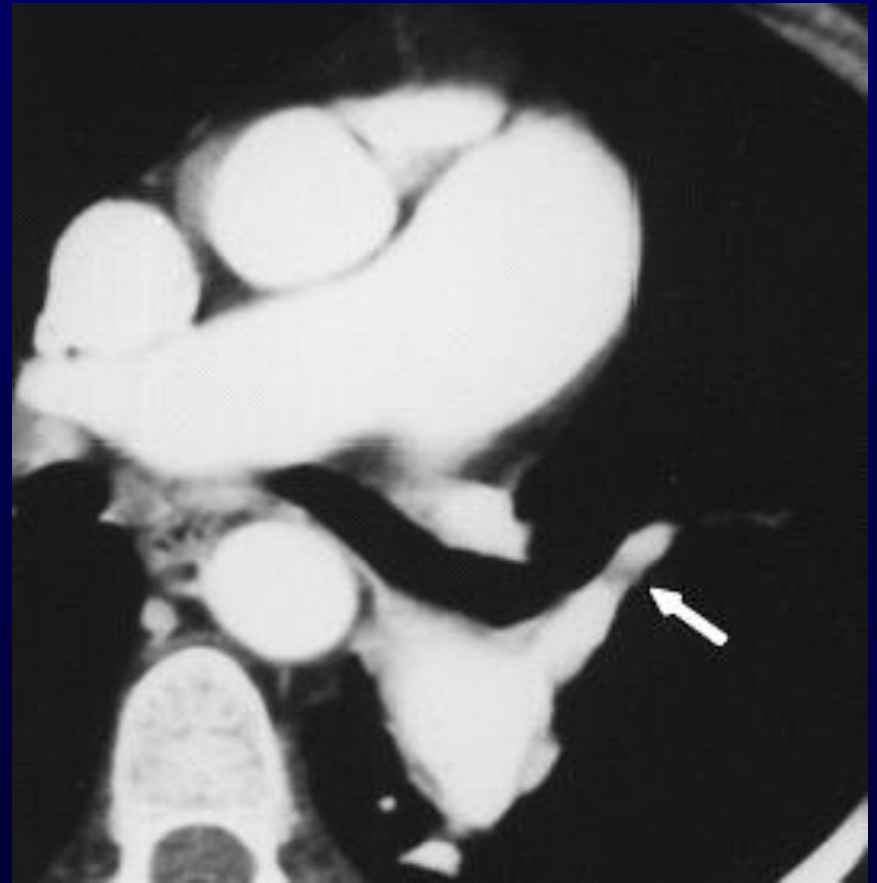
Ventilation–Perfusion Scintigraphy Is More Sensitive than Multidetector CTPA in Detecting Chronic Thromboembolic Pulmonary Disease as a Treatable Cause of Pulmonary Hypertension

Nina Tunariu¹, Simon J.R. Gibbs^{2,3}, Zarni Win⁴, Wendy Gin-Sing², Alison Graham¹, Philip Gishen¹, and Adil AL-Nahhas^{3,4}

- Retrospective: compared with DSA
- Of 78 CTEPH pts confirmed by DSA:
VQ: 75 high, 1 intermediate, 2 low
CTPA: 40 positive, 38 negative
- VQ: sens 96-97%, spec 90-95%
- CTPA: **sens 51%**, spec 99%

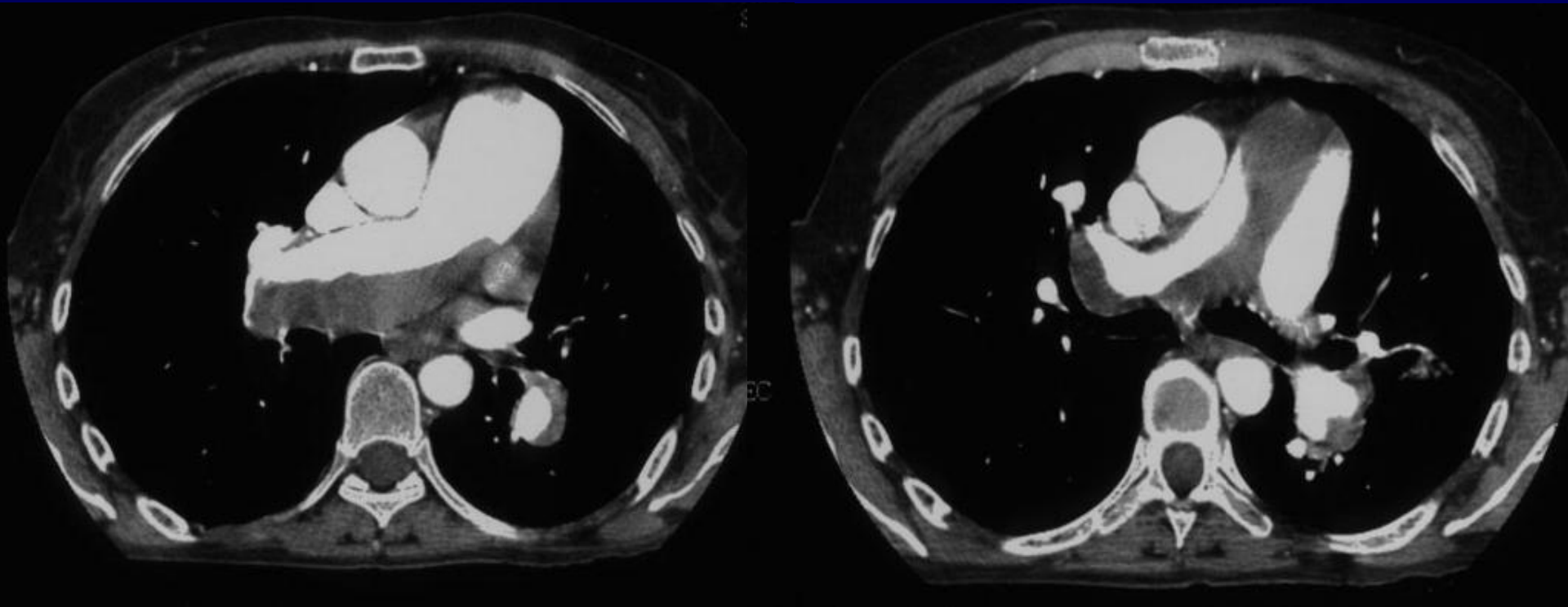
Pitfalls with CTPA:

(Under-Dx CTEPH, Over-Dx PAH?)

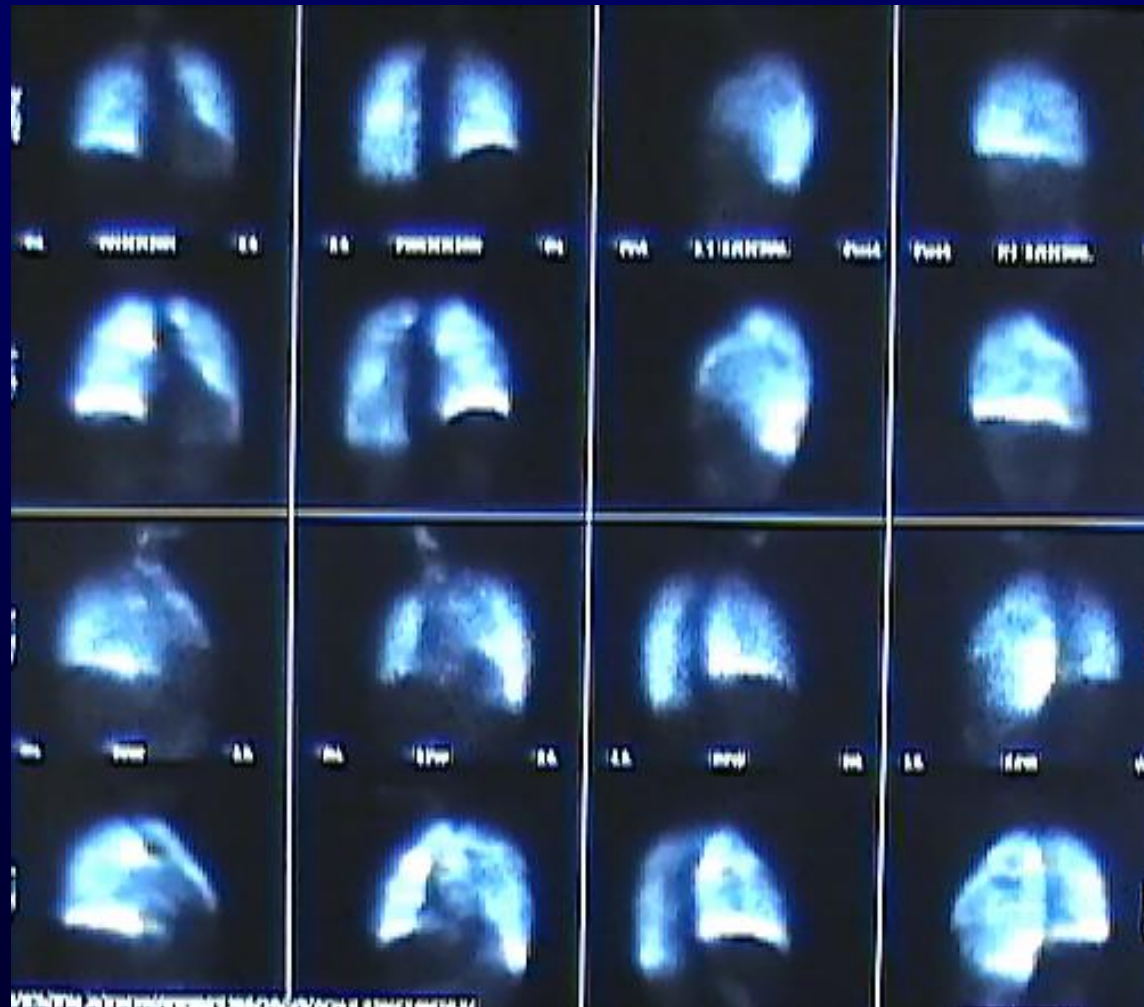


PAH-QuERI: PAH dx'd without VQ = 43%

CTPA: CTE disease?



VQ Scan (same patient)



CTEPH: Multi-Slice CTA



CTEPH: MR Angiogram

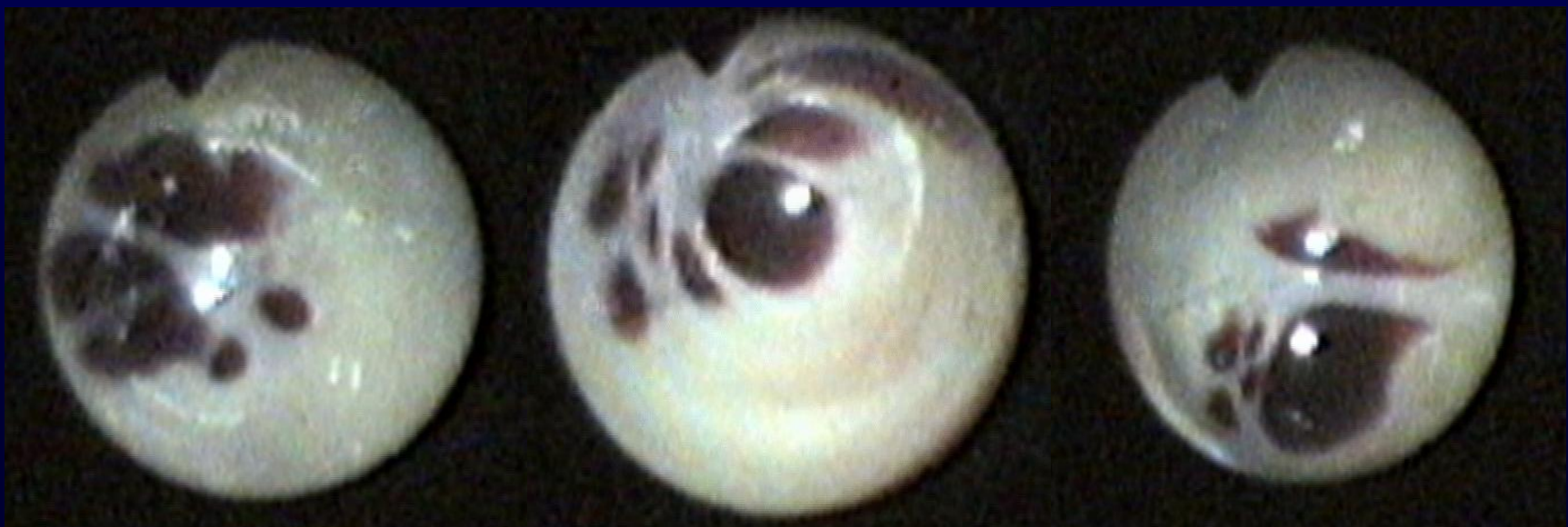
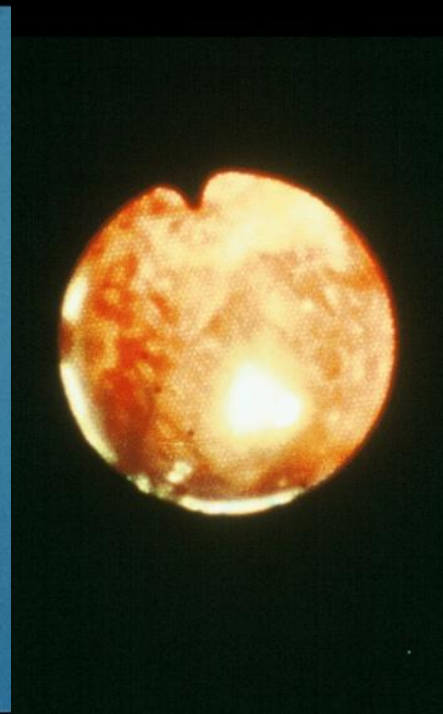


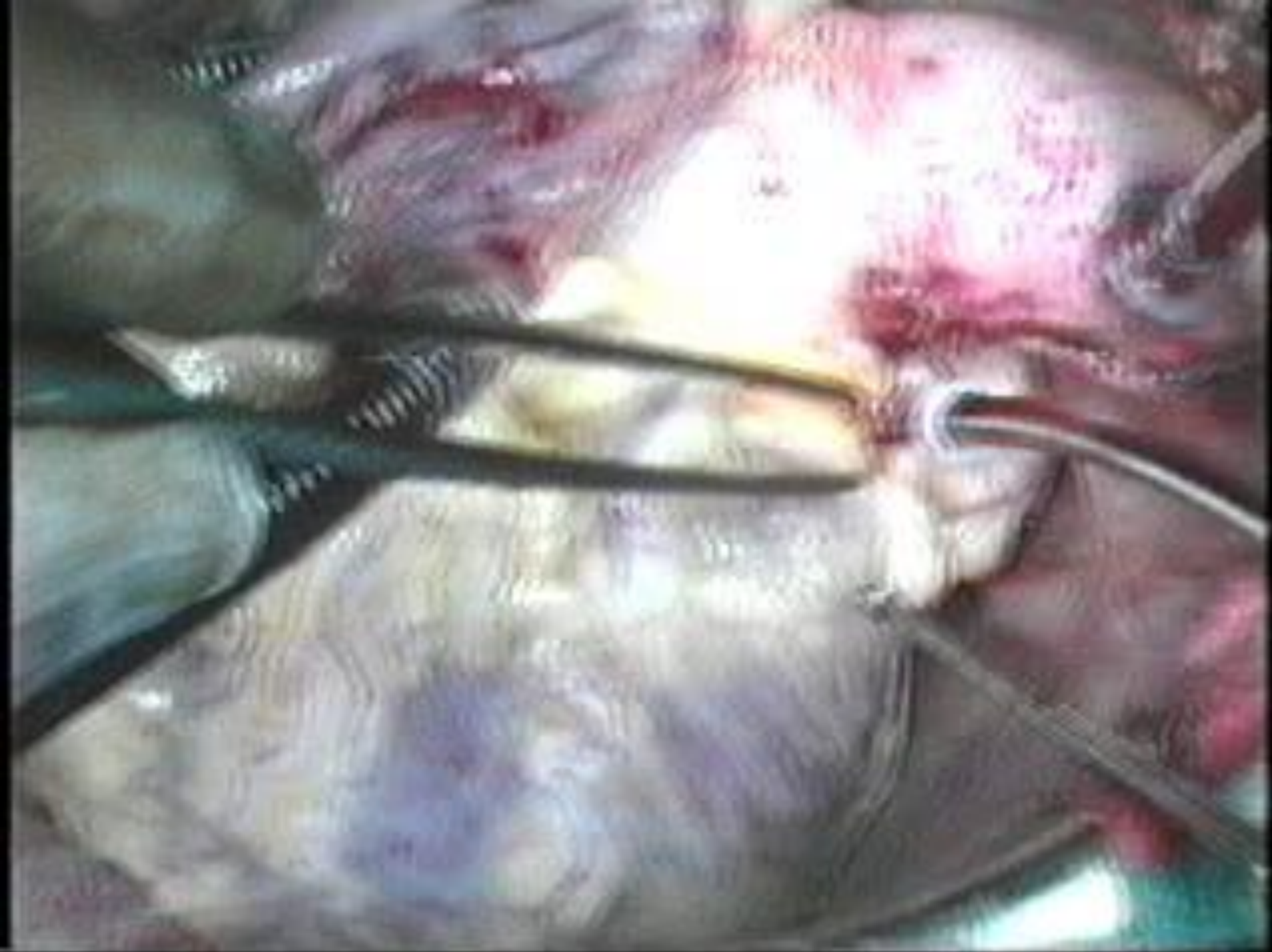
PRE



POST

CTEPH: Angioscopy

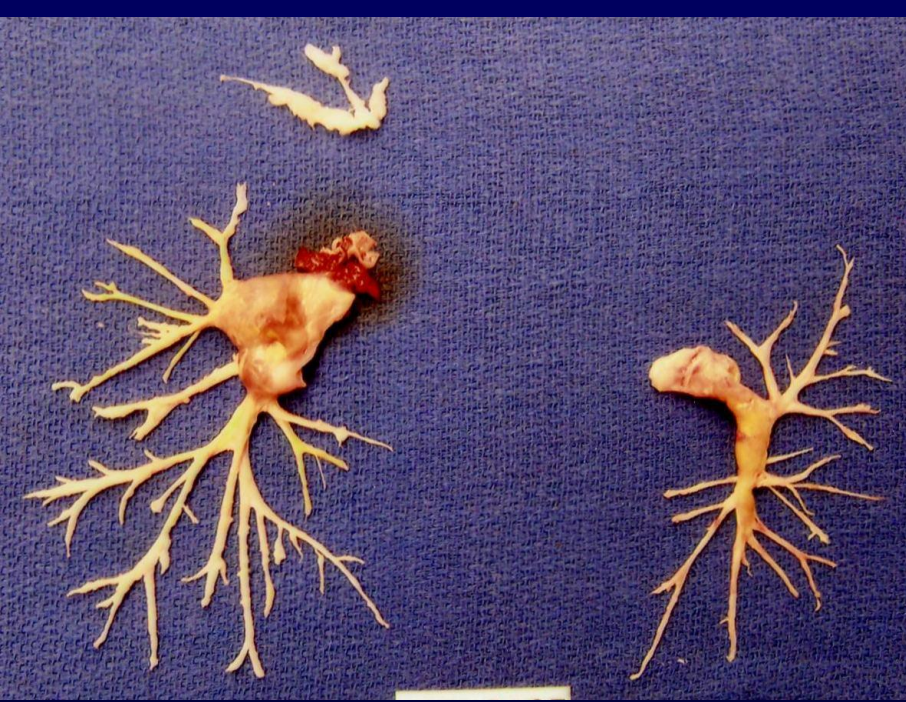




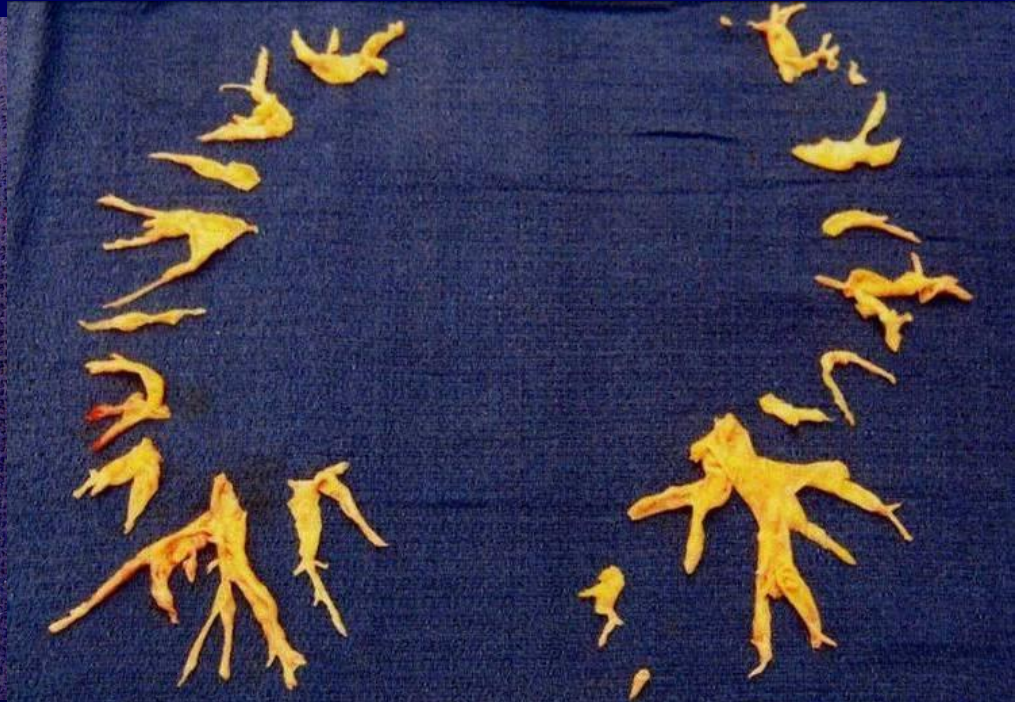
What's Impressive?



CTEPH: Experience Matters



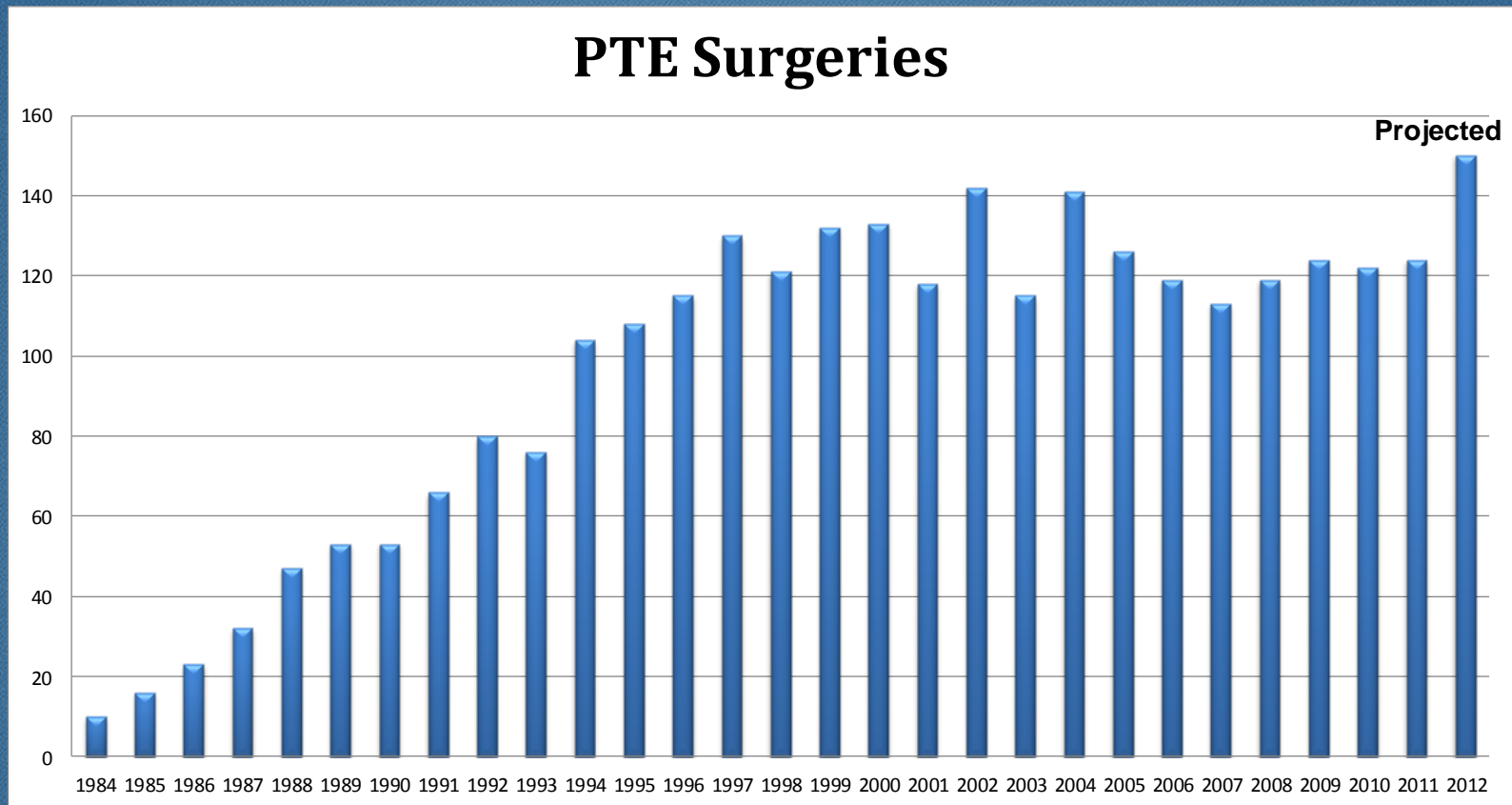
Thorough Endarterectomy



Jamieson Type III Disease

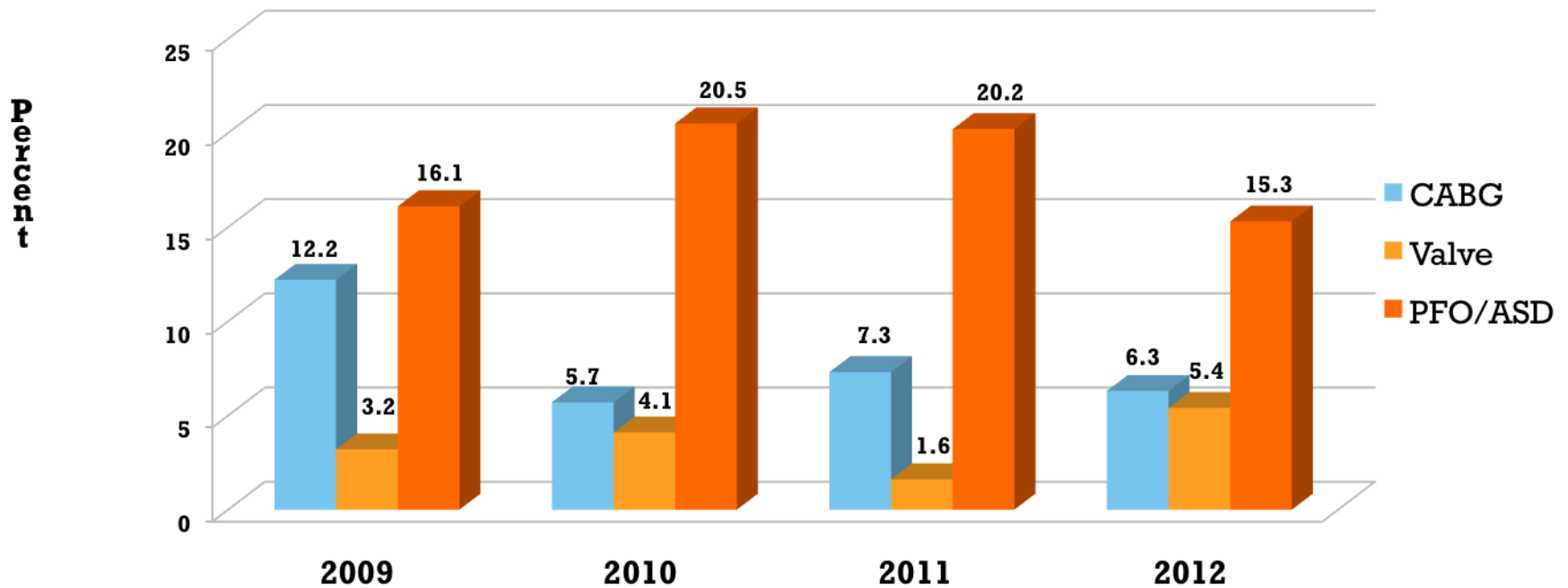
UCSD Surgical Experience

1984 - 2012

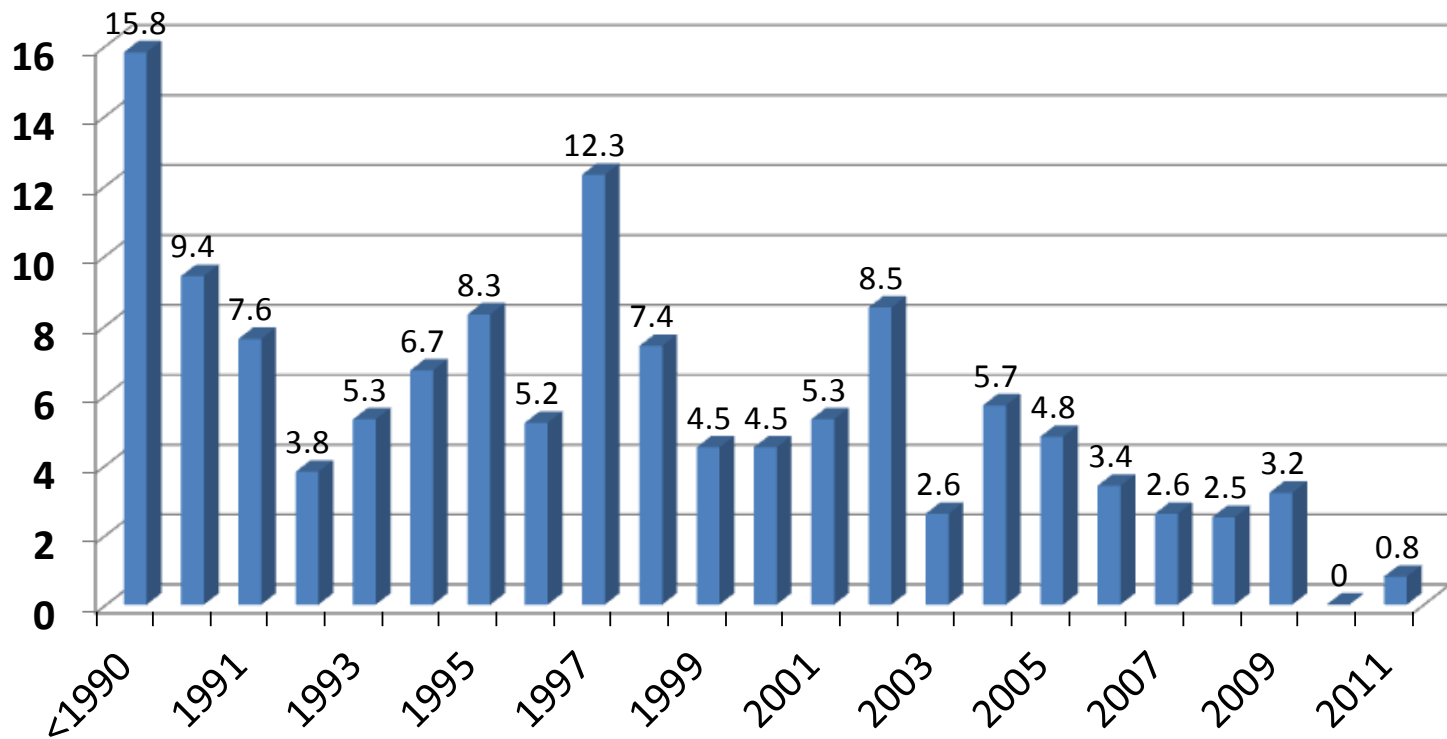


Not Always Just A PTE

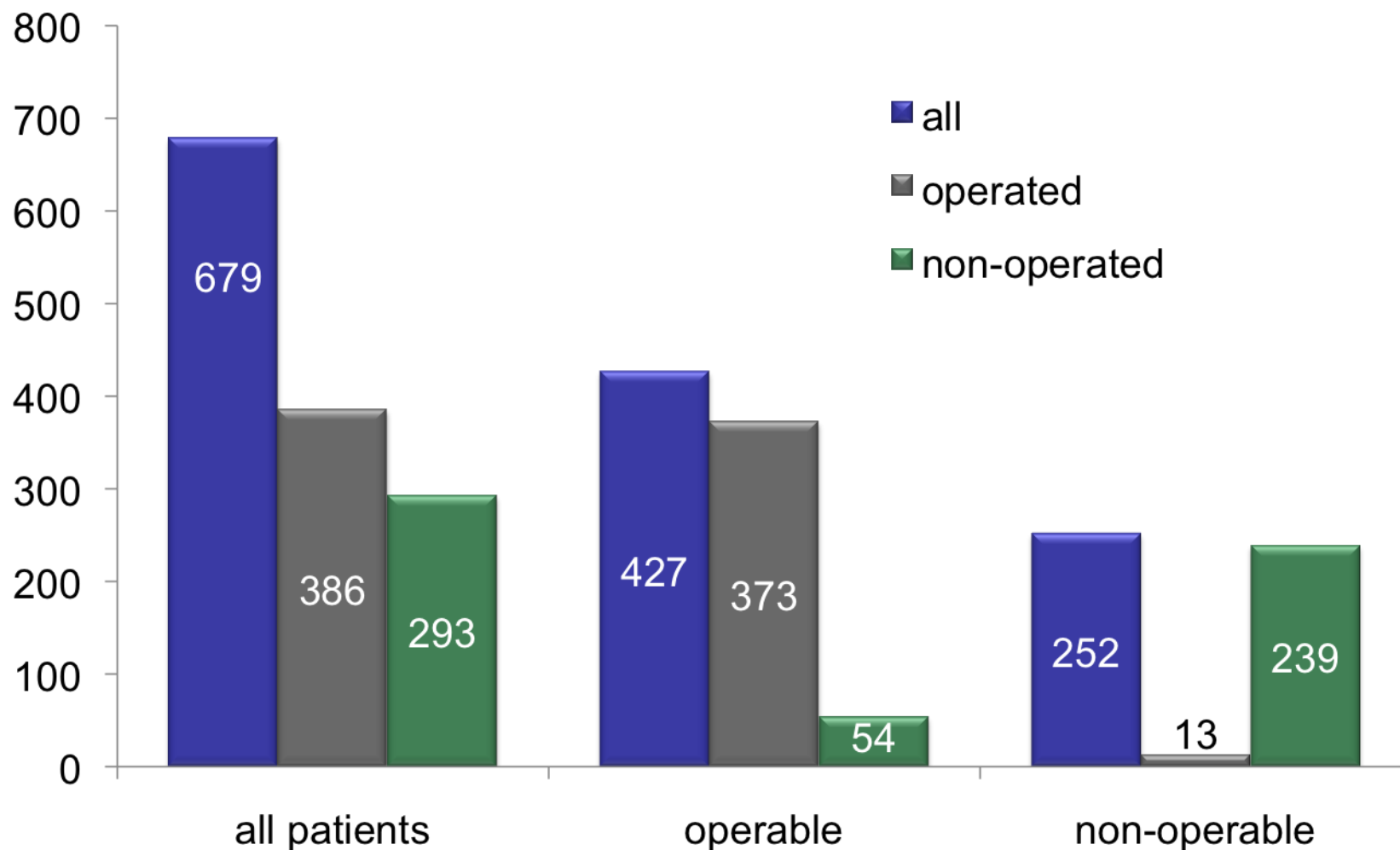
Other Surgical Procedures



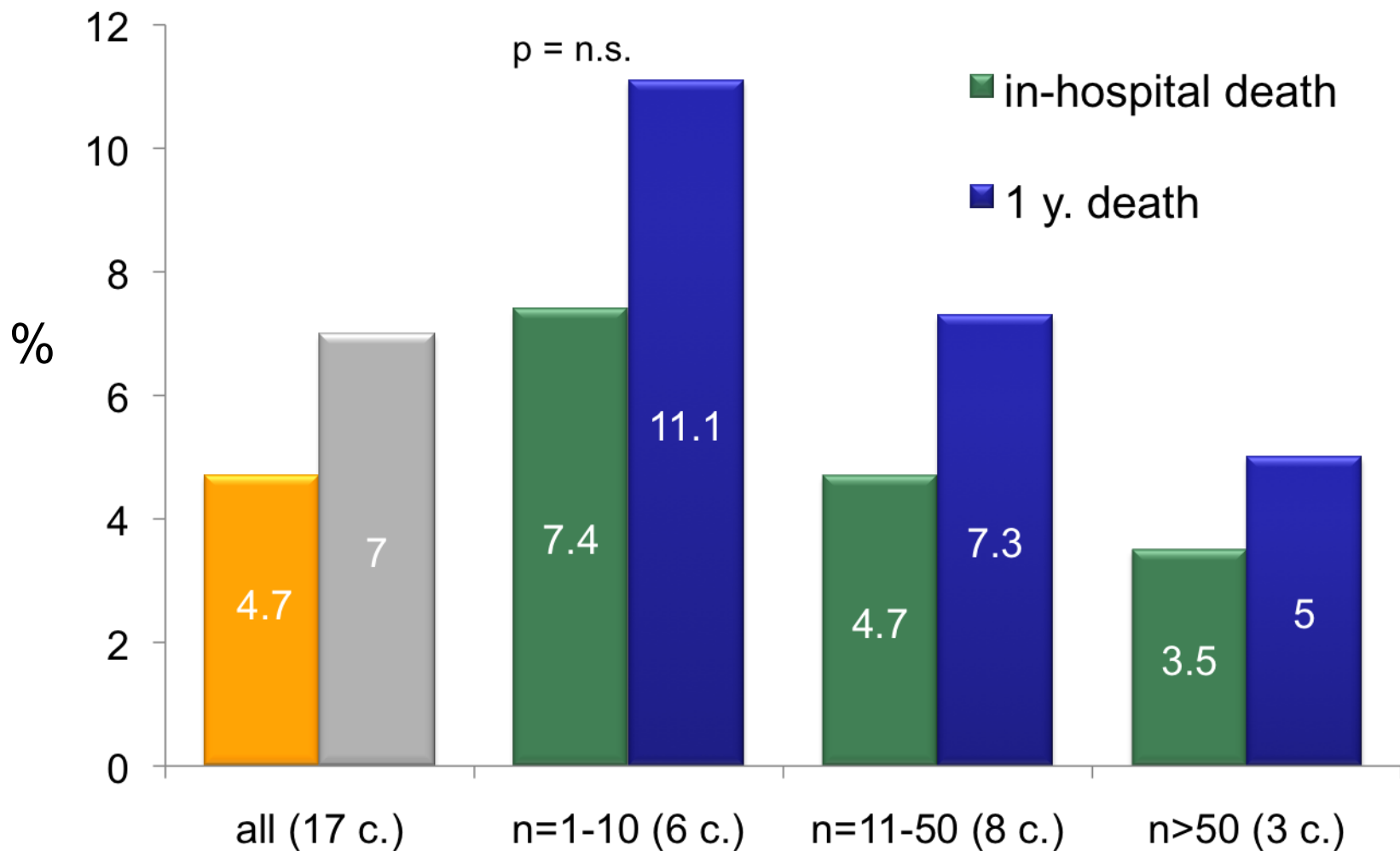
Perioperative Mortality Rates: UCSD



CTEPH Registry - Operability



Mortality – Center Expertise



International Scientific & Educational Workshop in CTEPH

27th & 28th June 2011

Robinson College, Cambridge, UK

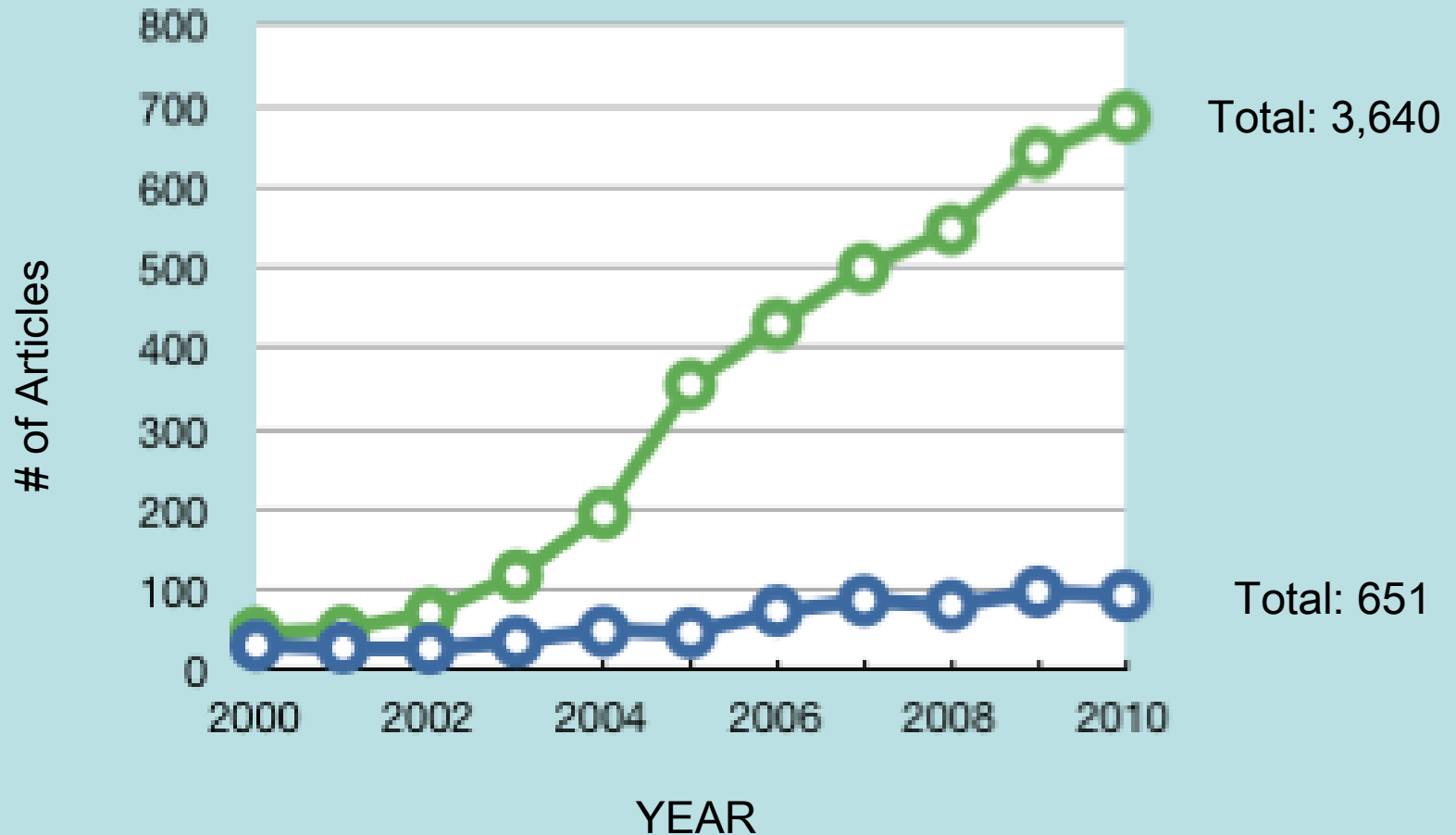
Papworth Hospital **NHS**
NHS Foundation Trust



The meeting has been organized by the Association for Research in CTEPH, in close collaboration with Papworth Hospital, a University of Cambridge Teaching Hospital

CTEPH (blue) vs PAH (green)

Pubmed results



CTEPH: Hot Topics

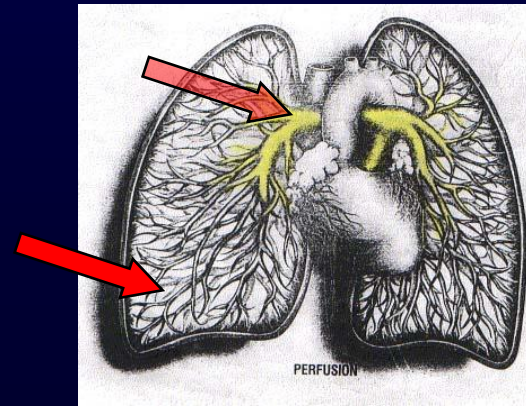
- **Operability Criteria**
- **Access/number of PEA Centers**
- **Role of Medical Therapy**

CTEPH and Operability Assessment

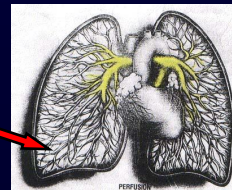
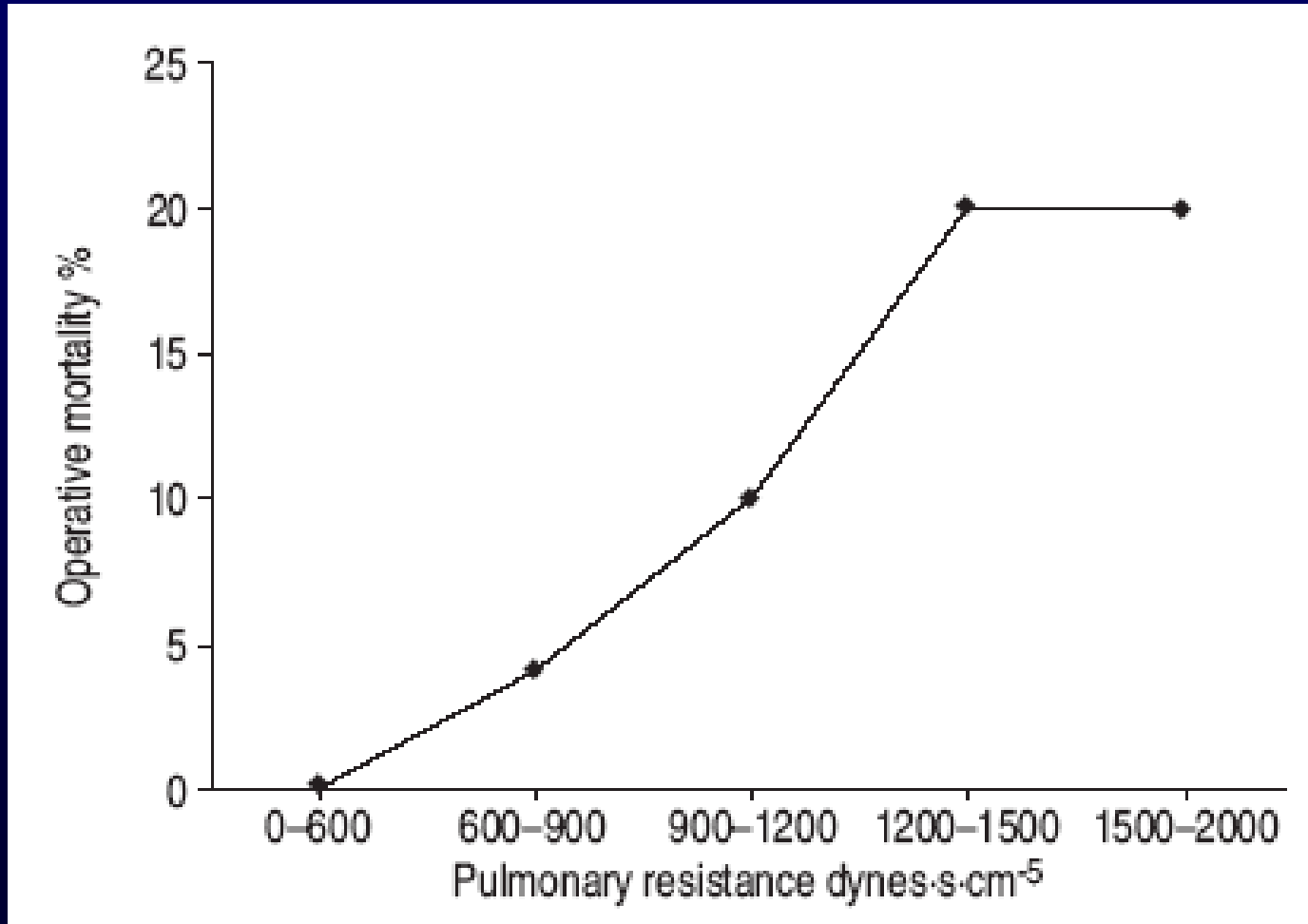
- 1) Is there chronic thromboembolic disease?
- 2) What is the PVR?
- 3) How experienced is your surgeon?
- 4) How experienced is your pre/post-operative team?

Preoperative Evaluation

- Is there CTE (proximal) disease?
 - VQ scan / PA angio / angioscopy
CT angio / MRA / PA ultrasound
- Is there microvascular (inoperable) disease?
 - Hemodynamic to radiographic discrepancy / Expert opinion



CTEPH: Preoperative PVR

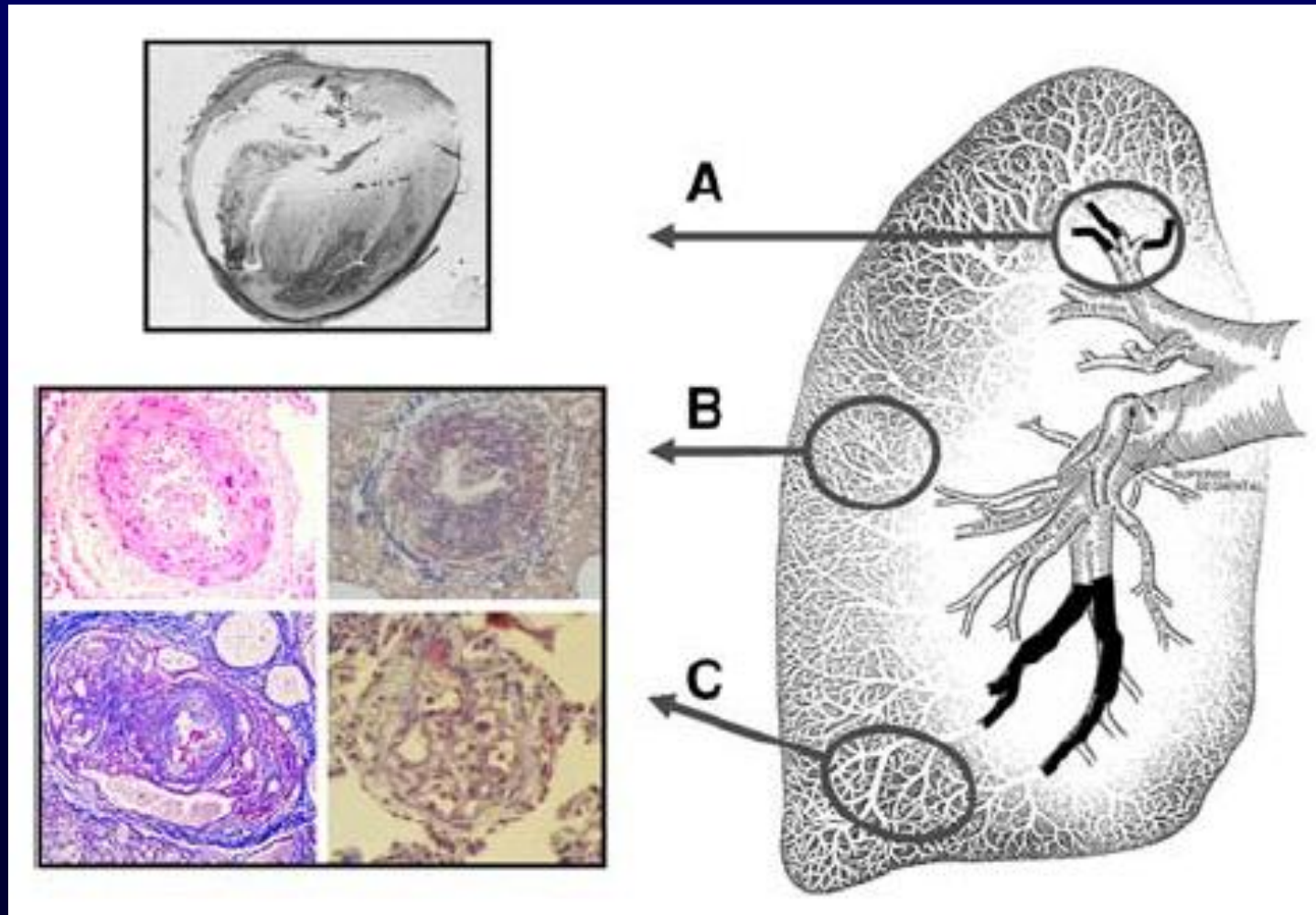


500 cases 1998 – 2002

Of 22 deaths (4.4%):

- 17 (77%) had residual pulmonary hypertension
- Post-op $PVR > 500 \text{ dsc}^{-5}$ had 30.6% mortality
- Post-op $PVR < 500 \text{ dsc}^{-5}$ had 0.9% mortality

Residual PH after PEA



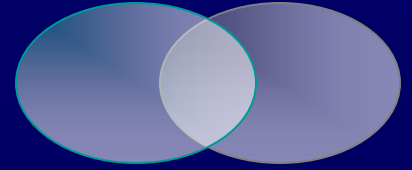
Rationale for Medical Therapy

- Pathologic evidence of concomitant small vessel disease (Moser & Bloor '93, Yi et al '00)
- Hemodynamic progression in the absence of new perfusion defects (Moser & Bloor '93)
- Discordance between hemodynamics and radiographically apparent burden of disease (Azarian R, et al. '97)
- Numerous uncontrolled reports of PAH-specific therapies having beneficial results in select CTEPH patients
- Availability of PAH therapy versus PEA center

“Inoperable” CTEPH: RCTs

- **Iloprost (AIR), n=57***
6MW+FC
Olschewski H, et al. *N Engl J Med* 2002; 347:322-9
- **Sildenafil (UK Pilot Study), n=19**
6MW
Suntharalingam J, et al. *Chest* 2008; 134:229-36
- **Bosentan (BENEFIT), n=157**
6MW or PVR
Jais X, et al. *JACC* 2008; 52:2127-34

CTEPH Summary



- **Defining role of medical treatment**
(to be continued...)
- **Screen PH with VQ scan**
- **Refer to PEA centers**
(not just for surgery but also for adjudication)
- **Always consider surgery**
(operability is dependent on numerous factors)