Identification and Evaluation of the Patient with Possible CTEPH

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DISCLOSURES:

- Co-Investigator: National CTEPH Registry (Bayer sponsored grant)
- Member, Advisory Board (uncompensated), CTEPH.com (Bayer sponsored)
Objective

Identify the best screening and diagnostic strategies for CTEPH
Question 1: What is the incidence of CTEPH following an acute pulmonary embolism?

A. 0%
B. 20-25%
C. 1-5%
D. 5-15%
Cumulative Incidence of CTEPH after a First Episode of Pulmonary Embolism without Prior Deep-Vein Thrombosis

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Patients (n=18)</th>
<th>Controls (n=287)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous PE</td>
<td>8 (44.4)</td>
<td>16 (5.6)</td>
</tr>
<tr>
<td>Idiopathic PE</td>
<td>10 (55.6)</td>
<td>103 (35.9)</td>
</tr>
<tr>
<td>Perfusion defect</td>
<td>62.6 ± 12.9</td>
<td>33.7 ± 18.6</td>
</tr>
</tbody>
</table>

Incidence of CTEPH after Acute Pulmonary Embolism

- Becattini P et al. Chest 2006;130:172-175 ➞ 0.8% of 259 patients
- Miniati M et al. Medicine 2006;85:253-262 ➞ 1.3% of 320 patients
- Dentali F et al. Thromb Res 2009; 124:256 ➞ 8.8% of 91 patients
- Klok F et al. Haematologica 2010; 95:970 ➞ 0.57-1.5% of 866 patients

Variability in patient selection, monitoring, confirmation of diagnosis

  146 patients, median follow up 26 months echo, CT at time of PE dx) CTEPH 4.8%
  Older, prior TE events, more proximal PEs
  At time of “acute PE” dx, higher PA pressures, 2 or more signs of CTED on CT
**Question 1:** What is the incidence of CTEPH following an acute pulmonary embolism?

A. 0%
B. 20-25%
C. 1-5%
D. 5-15%

**Answer:** C
Natural History of Chronic Thromboembolic Pulmonary Hypertension

- Genetic
- Medical Conditions (infection and inflammation)
- Prothrombotic Tendencies
- Recurrent TE Events/In-situ thrombosis

ACUTE PE | CTEPH

Small Vessel Changes
(Adaptive Vascular Remodeling – Lang IM)
Acute PE to CTEPH

Risk Factors: Medical Conditions

- Splenectomy
- Ventriculo-atrial shunts and infected PM
- Hx malignancy
- Thyroid replacement therapy
- Blood groups other than O

Comparative group: non thromboembolic PH
CTE Disease and CTEPH:

“Got to think of the diagnosis to make the diagnosis”

- High index of suspicion in patients who have unexplained exertional dyspnea
- Absence of a known history of venous thromboembolism does not exclude CTEPH dx
- Common Diagnostic errors: COPD, Asthma, Deconditioning....diagnostic delays
- Diagnosis of PH should be considered in all patients with unexplained dyspnea...and CTEPH in all patients with PH
Goals: Evaluation for suspected CTE Disease

• Confirm the diagnosis
• Establish proximal extent of disease

OPERABILITY ASSESSMENT
• Hemodynamic assessment
• Anticipated benefit
• Risk assessment: comorbidities

SURGICAL CANDIDACY
**Question 2:** The Ventilation-Perfusion scan in the pulmonary hypertensive patient is:

A. an “ancient” study and has no value  
B. less useful than a CT angiogram in assessing CTE disease  
C. is hard to interpret  
D. the recommended screening study for CTEPH
Diagnostic Approach to Patients with suspected CTE Disease

Hoeper et al. Am Coll Cardiol 2009; 54:S85-S96
Perfusion Scans and PAH

Pulmonary Arterial Hypertension Quality Enhancement Research Initiative Registry

43% of PAH patients: No V/Q during the evaluation process

CTEPH: Screening V/Q scan

<table>
<thead>
<tr>
<th>Indicator</th>
<th>V/Q (1)*</th>
<th>V/Q (2)†</th>
<th>CTPA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity (%)</td>
<td>97.4</td>
<td>96.2</td>
<td>51.3</td>
</tr>
<tr>
<td>Specificity (%)</td>
<td>90</td>
<td>94.6</td>
<td>98.3</td>
</tr>
<tr>
<td>Accuracy (%)</td>
<td>92.5</td>
<td>95.2</td>
<td>82.8</td>
</tr>
<tr>
<td>NPV (%)</td>
<td>98.5</td>
<td>97.9</td>
<td>79.7</td>
</tr>
<tr>
<td>PPV (%)</td>
<td>83.5</td>
<td>90.3</td>
<td>97.6</td>
</tr>
</tbody>
</table>

*Intermediate with high-probability scans as indicative of CTEPH. †Only high-probability scans as indicative of CTEPH.

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Answer: D
Perfusion Scans

PAH

CTEPH

Segmental CTEPH

Pulmonary Arteritis

Sarcoma
CTEPH: CTA Findings

Acute to Chronic
CTEPH: CTA Findings

Eccentric thrombus

Lining thrombus, Enlarged main PA (PH)

Bronchial arterial collateral vessels

Mosaic Perfusion
Competing Diagnoses: CT Findings

Fibrosing Mediastinitis

Parenchymal Lung Disease

Sarcoma
CTA: Segmental level CTE Disease
CTEPH: Pulmonary Angiography

◆ The ‘gold standard’
◆ Essential (in most cases) to establish operability for those patients with more distal CTE disease

- Safe -- if due precautions taken
- Single injection, right and left PA
- Modification of non-ionic contrast volume / rate
- Careful patient monitoring, O2, etc.

► Biplane study

- “Pouch” defects
- Webs and bands
- Intimal irregularity
- Abrupt vascular narrowing
- Complete obstruction at main, lobar or segmental levels
Pulmonary Angiogram: Mapping
CTEPH and Pulmonary Angiography

• Hemodynamic assessment at time of procedure
• Complete assessment process for surgical candidacy
MRI & Dual Energy CT

MRI

- To evaluate Hemodynamics (RV function) and response to therapy
- To evaluate clot burden with MRA angiogram and assess functional significance by perfusion

Dual Energy CT

- Same angiographic assessment as standard CT but also provides functional impact by assessing parenchymal perfusion
Final Thoughts...The “pearls”

• CTEPH needs to be considered in all PH patients
• V/Q scan is the recommended screening study in the evaluation for CTEPH
• Pulmonary angiography and CT (MR) provide complementary information
• Pulmonary angiography: “gold standard” in evaluation of segmental/subsegmental CTED
• Centers with expertise in CTEPH should be contacted to assist in operability assessment