Chronic thromboembolic pulmonary hypertension, or CTEPH, is defined as mean pulmonary arterial pressure (mPAP) of at least 25 mmHg and pulmonary capillary wedge pressure (PCWP) of no more than 15 mmHg in the presence of multiple chronic or organized occlusive thrombi or emboli in the elastic pulmonary arteries (the main, lobar, segmental, subsegmental) after at least 3 months of effective anticoagulation. CTEPH is a complex and misunderstood disease, and numerous myths about CTEPH and its treatment persist. Here, we present evidence that should help dispel some commonly held myths about the diagnosis of CTEPH and about pulmonary thromboendarterectomy (PTE) surgery, the only potentially curative treatment for CTEPH.

**CTEPH Definition**

**MYTH:**
If a patient recovers from an acute pulmonary embolism, she won’t go on to develop CTEPH.

**Evidence:** While the likelihood of developing CTEPH after an acute pulmonary embolism (PE) is small, it is not negligible. CTEPH is a rare complication of a common disease.

**Diagnosis**

- A prospective follow-up study by Pengo et al of 314 consecutive patients found that 3.8% eventually developed CTEPH within 2 years of their first symptomatic episode of acute PE

- There are several factors that increase the risk that CTEPH will eventually develop subsequent to an acute PE. These include:
  - Large perfusion defects
  - Recurrent or idiopathic PE
  - Young or old age
  - Persistent PH 6 months after the embolism has occurred
  - Systolic PAP >50 mmHg at initial manifestation of PE

  Recognizing this risk, the ACCF/AHA 2009 expert consensus document on pulmonary hypertension (PH) recommends that a ventilation/perfusion (V/Q) scan be performed in patients who show symptoms of PH 3 months after having an acute PE.

- Studies report that about a quarter to as many as 63% of CTEPH patients may present without a history of acute PE or deep vein thrombosis.

MYTH: The V/Q is unnecessary in the evaluation of pulmonary hypertension.

**Evidence:** Multiple PH guidelines—including those from the Fifth World Symposium on Pulmonary Hypertension (WSPH) CTEPH Task Force—recommend that the V/Q scan be considered an essential CTEPH screening tool as part of the PH diagnostic algorithm.

The diagram below communicates a mnemonic, SCAR, developed by the WSPH CTEPH Task Force. Along with echocardiography, the V/Q scan provides information that leads to a suspicion of CTEPH, but it is only with right heart catheterization (RHC) and pulmonary angiography that a diagnosis of CTEPH can be confirmed.

- Computed tomographic pulmonary angiography (CTPA) may be used to provide complementary diagnostic information, but it does not replace the V/Q scan as a screening test
  - In a retrospective study of 227 patients, Tunariu et al reported CTPA had a sensitivity for detecting CTEPH of 51%, while the V/Q scan had a sensitivity >96%
  - The WSPH noted that use of CTPA as a screening tool instead of V/Q scanning, “may lead to potential misdiagnosis of PAH and underdiagnosis of CTEPH”
- The V/Q scan is the preferred and recommended method of screening for CTEPH
  - Every patient being evaluated for PH should undergo a V/Q scan if CTEPH has not already been excluded by a prior normal V/Q scan


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Assessing Operability For PTE Surgery

**MYTH:**
PTE surgery cannot be safely performed in patients who have more severe PH or who are elderly, obese, or otherwise very sick.

**Evidence:** There are no absolute contraindications to PTE surgery based on severity of pulmonary hypertension, age, body mass index, or presence of comorbidities.

- Madani and colleagues from University of California, San Diego (UCSD) reported a series of 500 patients who had PTE surgery and found that among the patients who had preoperative pulmonary vascular resistance (PVR) >1000 dynes, which indicates comparatively severe pulmonary hypertension, overall mortality was 4.1% vs 1.6% in patients who had preoperative PVR <1000.


- A recent analysis of data collected by researchers at UCSD evaluated outcomes after PTE for CTEPH in a cohort that included 120 patients who were obese (body mass index [BMI] >30) and 208 patients who were not obese (BMI<30) demonstrated no significant difference in mortality (2.4% for obese vs 0.8% for nonobese patients) or post-PTE hemodynamics between the groups.


- Patients who have underlying cardiovascular disease may be able to undergo corrective procedures—eg, valve replacement or coronary artery bypass grafting—at the same time as PTE.

- Those with hepatic or renal insufficiency at baseline may have complications during the perioperative period, but in the long term, hepatic and renal function may improve along with the general improvements in CTEPH following PTE surgery.


Any major surgery may pose a greater risk for older patients as compared with younger patients. Similarly, obesity, serious comorbidities, and severity of PH certainly need to be considered by the experienced CTEPH team assessing a patient’s operability for PTE surgery. However, none of these factors automatically disqualifies patients from potentially curative PTE surgery.

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**Comparable mortality rates in older and younger patients (1/2006-3/2011)**

<table>
<thead>
<tr>
<th>Time Frame</th>
<th>&lt;70 years, n=308</th>
<th>≥70 years, n=103</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>In hospital</td>
<td>4.6%</td>
<td>7.8%</td>
<td>.21</td>
</tr>
<tr>
<td>1 year</td>
<td>8.6%</td>
<td>14.1%</td>
<td></td>
</tr>
<tr>
<td>2 year</td>
<td>10.1%</td>
<td>15.9%</td>
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<tr>
<td>3 year</td>
<td>12.3%</td>
<td>15.9%</td>
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</tr>
</tbody>
</table>

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PTE Surgery

**MYTH:**

PTE surgery is still an experimental and dangerous procedure.

**Evidence:** PTE surgery is not experimental. Rather, it’s a well-established, reproducible procedure that represents the only potential cure for patients who have CTEPH.

- The PTE surgical technique was developed and first performed in 1970.
  - The blue table documents the PTE history of a single experienced center. Mortality rates have consistently decreased, such that none of their last 260 PTEs, as reported by Madani et al in 2012, resulted in death.
  - At the most experienced centers, 96.5% survive to discharge.


- Since its inception over 4 decades ago, the safety of PTE surgery has improved markedly, such that experienced centers have been able to achieve in-hospital mortality rates of <5%.


**MYTH:**

Drug therapy can be used in place of PTE surgery.

**Evidence:** According to the WSPH CTEPH Task Force, drug therapy is no substitute for PTE in a CTEPH patient who has operable disease.

- PTE is the standard and recommended treatment for CTEPH, and it is the only treatment option that can potentially cure CTEPH.
- Guidance from the WSPH makes it clear that all patients with CTEPH should be assessed for operability, and those who have operable disease should be referred for PTE without delay.
- Furthermore, the role of “bridging” with drug therapy in CTEPH has not been sufficiently studied and should be reserved for controlled investigation.


To learn more about CTEPH, please visit CTEPH.com

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