

# Group 4 Pulmonary Hypertension

## Chronic Thromboembolic Pulmonary Hypertension: Epidemiology, Pathophysiology, and Treatment

Nick H. Kim, MD

### KEYWORDS

- Chronic thromboembolic pulmonary hypertension • Chronic thromboembolism
- Pulmonary embolism • Pulmonary thromboendarterectomy • Pulmonary endarterectomy
- Riociguat • Balloon pulmonary angioplasty

### KEY POINTS

- Pulmonary embolism history can be absent in chronic thromboembolic pulmonary hypertension (CTEPH).
- Negative computed tomography pulmonary angiogram does not rule out CTEPH.
- Pulmonary thromboendarterectomy remains the treatment of choice for CTEPH.
- Operability assessment should be performed by an experienced CTEPH team.

The first successful pulmonary thromboendarterectomy (PTE) for the treatment of chronic thromboembolic pulmonary hypertension (CTEPH) was reported nearly 20 years before the introduction of both heart-lung transplantation and intravenous epoprostenol for the treatment of pulmonary arterial hypertension (PAH).<sup>1–3</sup> Fast forward another 30 years, and much has evolved in our understanding and treatment of pulmonary hypertension (PH), and with that, so has our approach to the diagnosis and management of patients with CTEPH.

### EPIDEMIOLOGY

CTEPH is a complication of pulmonary embolism.<sup>4,5</sup> Although the true incidence of CTEPH following acute pulmonary embolism remains unknown, reports have ranged widely from 0.4% to 9.1%.<sup>4,6–10</sup> The variability of the incidence reports

may reflect differences in patient selection and methodology across these reports. Whether these rates represent true incidence of CTEPH after acute pulmonary embolism, or combination of incident and prevalent cases remains speculative. For example, in the series of Pengo and colleagues,<sup>4</sup> no additional CTEPH was detected beyond 2 years from the initial acute pulmonary embolism, and one of the cases reportedly developed near-systemic PH within just 5 months from the acute event. The characteristics of these cases arguably raise concerns that the series of Pengo and colleagues<sup>4</sup> may not be solely an incident series of CTEPH occurring after acute pulmonary embolism, but rather one that unintentionally included both incident and previously unrecognized CTEPH cases.

Additional efforts are currently under way to prospectively capture incident cases of CTEPH by screening after acute, first-time pulmonary

---

Disclosures: Consultancy/Speakers Bureau: Actelion, Bayer; Board Member: CTEPH.com.  
Division of Pulmonary and Critical Care Medicine, University of California, San Diego, 9300 Campus Point Drive, MC 7381, La Jolla, CA 92037, USA  
E-mail address: [h33kim@ucsd.edu](mailto:h33kim@ucsd.edu)

Cardiol Clin 34 (2016) 435–441

<http://dx.doi.org/10.1016/j.ccl.2016.04.011>

0733-8651/16/\$ – see front matter © 2016 Elsevier Inc. All rights reserved.

embolism cases to better assess the true incidence of CTEPH. Nevertheless, any measurement or estimate of CTEPH incidence after acute pulmonary embolism may underestimate the overall burden of CTEPH because as many as 25% to 30% of patients with CTEPH are diagnosed without a prior clinical history of pulmonary embolism, and nearly half of these patients may not have a history of deep venous thrombosis.<sup>11,12</sup> Therefore, a key take-home point when evaluating patients presenting with PH is that the lack of a venous thromboembolism history should not exclude CTEPH as a possibility.

**Table 1** shows epidemiologic data used to estimate the incidence of CTEPH to better appreciate the scope of CTEPH in the United States.<sup>13</sup> The calculations shown are based on the CTEPH incidence rate reported from Klok and colleagues.<sup>7</sup> This study included the largest pulmonary embolism series (866 consecutive cases) screened for CTEPH following standard diagnostic guidelines including right heart catheterization. Even using this lower range of the reported incidence rates after pulmonary embolism, approximately 3400 cases of CTEPH in the United States might be expected each year. Combined with the approximately 30% of CTEPH cases operated without a history of prior pulmonary embolism, the overall estimate of new CTEPH cases diagnosed in the United States each year jumps to nearly 5000 new CTEPH cases per year.

An important question today then is: Where are these patients with CTEPH and how are they being treated? Although precise data on the number of PTE surgeries performed in the United States is not known, estimated 300 to 400 PTEs are being performed in the United States per year today. However, based on estimates from **Table 1**, this represents fewer than 8% of incident CTEPH

**Table 1**  
Estimated annual incidence of CTEPH in the United States

	Estimates
PE cases per year <sup>13</sup>	600,000
Incidence of CTEPH after PE <sup>7</sup>	0.57%
CTEPH cases after PE per year	3420
Additional 30% of CTEPH cases per year without prior pulmonary embolism	1466
Total overall CTEPH cases estimated per year	4886

**Abbreviations:** CTEPH, chronic thromboembolic pulmonary hypertension; PE, pulmonary embolism.

cases being operated annually. Even when accounting for the limitations of such extrapolated estimates, there appears to be a large discrepancy between number of potentially surgically treatable cases and surgically treated volume. **Table 2** lists the plausible reasons behind this large gap. In addition to possibly inaccurate incidence rates, cases of CTEPH may not be properly diagnosed, and if diagnosed, may not be referred for surgical treatment as recommended consistently by best practice guidelines.<sup>14,15</sup>

One particular diagnostic pitfall in CTEPH is the failure to obtain a lung ventilation perfusion (VQ) scan to screen for CTEPH during the workup of PH.<sup>16</sup> Computed tomography (CT) pulmonary angiography is often (erroneously) used in place of the VQ scan. Unfortunately, CT pulmonary angiography lacks adequate sensitivity to detect

**Table 2**  
Potential explanations for the gap between estimated CTEPH incidence from Table 1, and the relatively small number of PTE cases performed in the United States

Explanation	Result
<i>Assumptions are incorrect</i>	
Examples:	Error in true incidence of CTEPH
<ul style="list-style-type: none"> <li>• PE estimate may not be accurate</li> <li>• Some PE cases may have had CTEPH already (ie, not incident PE)</li> <li>• CTEPH true incidence may be lower</li> </ul>	
<i>CTEPH is not being diagnosed</i>	
Examples:	CTEPH is underestimated
<ul style="list-style-type: none"> <li>• Failure to recognize/diagnose CTEPH</li> <li>• CTEPH Incorrectly diagnosed as PAH</li> <li>• CTEPH Incorrectly diagnosed as recurrent pulmonary embolism</li> </ul>	
<i>Under referral for CTEPH evaluation/treatment</i>	
Examples:	CTEPH is underestimated
<ul style="list-style-type: none"> <li>• Providers not aware of PTE surgery or have limited access</li> <li>• Operability is being decided locally</li> <li>• Medical treatment is elected instead of PTE</li> </ul>	

**Abbreviations:** CTEPH, chronic thromboembolic pulmonary hypertension; PAH, pulmonary arterial hypertension; PE, pulmonary embolism; PTE, pulmonary thromboendarterectomy.

Download English Version:

<https://daneshyari.com/en/article/2897719>

Download Persian Version:

<https://daneshyari.com/article/2897719>

[Daneshyari.com](https://daneshyari.com)