10 Chronic Thromboembolic Pulmonary Hypertension

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Abstract

Chronic thromboembolic pulmonary hypertension (CTEPH) is an important form of pulmonary hypertension to detect because prompt treatment can lead to a surgical cure. The true incidence is unknown, but it is estimated to occur in 1% to 3% of patients following acute thromboembolism. Detection may be difficult, because symptoms are nonspecific and other diagnoses are often made before that of CTEPH is entertained. Routinely screening all pulmonary hypertension

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patients with a ventilation–perfusion scan will detect most, however. Candidates for thromboendarterectomy are evaluated using right heart catheterization, computerized tomographic angiography, and pulmonary angiography, seeking those with proximal obstructions that can be removed surgically. Patients who are not candidates for thromboendarterectomy because of comorbidities, very high pulmonary vascular resistances, or mainly distal disease may still receive medical therapy or be considered for lung transplantation.

**Key Words:** chronic thromboembolic pulmonary hypertension; thromboendarterectomy; lung transplantation; thromboembolism; pulmonary angiography; pulmonary angioscopy.

### 1. INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) represents a unique cause of secondary pulmonary hypertension by virtue of its potentially remedial nature. Therefore, it is imperative to identify patients with this disorder. Unlike most other variants of pulmonary hypertension in which modest improvements in functional status and clinical stability are the usual achievable therapeutic endpoints, thromboembolic pulmonary hypertension is potentially curable.

The first surgical procedure for CTEPH was performed in 1958 (1,2). Over the next 26 years, only 85 patients who had undergone thromboendarterectomy were reported in the medical literature, with a mortality of 22% (3). Since 1985, the number of pulmonary thromboendarterectomies has increased dramatically, the majority (over 2,000) having been performed at the University of California at San Diego. Other centers have begun to perform the procedure, including those in Canada, Great Britain, France, Austria, Germany, Italy, Australia, and the Netherlands. The overall surgical mortality has declined to the range of 4% to 8% as established programs have enhanced their skills with the preoperative assessment, surgical management, and perioperative care of patients suffering from this disease (4–6). As is the case with other highly complex surgical procedures requiring a multidisciplinary approach, mortality seems to be related to the surgical volume (7–9).

Our understanding of the epidemiology of CTEPH is evolving. Based on the number of patients referred for thromboendarterectomy, it was originally thought that 0.1% to 1% of patients who suffered from pulmonary embolism would subsequently develop pulmonary hypertension (7). However, more recent data suggest that 0.8% to as many as 3.1% of patients may develop CTEPH after an initial episode of pulmonary embolism (10,91), with an even higher occurrence after