Operability assessment in chronic thromboembolic pulmonary hypertension (CTEPH): Don't miss the chance of a second opinion!

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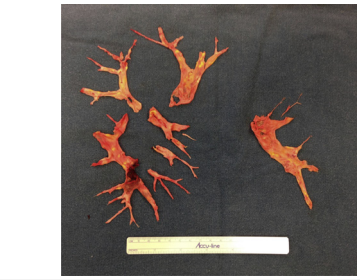

Good outcome is determined by a correct diagnosis and not the extent of disease on imaging.

Central Message

Pulmonary endarterectomy is the mainstay of therapy for chronic thromboembolic pulmonary hypertension (CTEPH) and accessibility to a second surgical opinion from experienced centers is a crucial component in the evaluation of patients with CTEPH.

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Jenkins and colleagues[1] present the impact that a central and local adjudication committee had on patient selection for pulmonary endarterectomy in the CHEST-1 (Chronic Thromboembolic Pulmonary Hypertension Soluble Guanylate Cyclase Stimulator Trial-1) study. They observed that of the 312 patients initially considered inoperable, 69 (22%) were determined to be surgical candidates after central (48 of 188 patients, 25%) or local (21 of 124 patients, 16%) review. These findings highlight 2 major issues among patients with chronic thromboembolic pulmonary hypertension (CTEPH): (1) misdiagnosis and (2) operability. Patients with segmental and subsegmental disease are surgical candidates with excellent early- and long-term outcome in experienced centers[2,3]; however, these patients can easily be misdiagnosed if the imaging is not optimal or mislabeled as inoperable in centers with less experience.

The diagnosis of CTEPH relies on ventilation–perfusion (VQ) scan as the first step to demonstrate the presence of mismatched perfusion defects. Pulmonary angiography is then performed to confirm the presence of chronic thromboembolic disease. Interestingly, 15% of the patients reviewed by the local or central adjudication committee did not have a VQ scan. VQ scan remains a key diagnostic step and a very valuable imaging in the assessment of patients with segmental and subsegmental disease to ensure that the correct diagnosis is made before surgery.

The authors demonstrate in this paper the importance of re-evaluating operability by a second experienced center in the event a patient is deemed inoperable. This finding is in agreement with previous recommendation made during the fifth world symposium on pulmonary hypertension.[4] This step is important, because pulmonary endarterectomy remains the mainstay of therapy for CTEPH, with an operative mortality of less than 5%, a 10-year survival of 75% or greater, and excellent quality of life.[3,5,6]

The authors, unfortunately, did not review the reasons to determine operability and inoperability in this series of patients. They point out that the assessment is subjective and depends on the experience of the surgeon. With the avenue of balloon pulmonary angioplasty, however, a more objective assessment and a clear classification system to describe the location of the thromboembolic material will be important to be able to determine the role of this new treatment option in the management of patients with CTEPH.

The authors discuss the correlation between the extent of thromboembolic disease and the severity of the pulmonary hypertension in the decision making for surgery. This correlation, however, is extremely subjective with no clear definition and, in our experience, has been abandoned from the algorithm to determine surgical candidacy.[7] The amount of material found intraoperatively is underestimated by the radiological evaluation, particularly in patients with segmental and subsegmental disease, and thus the correlation is very difficult to predict

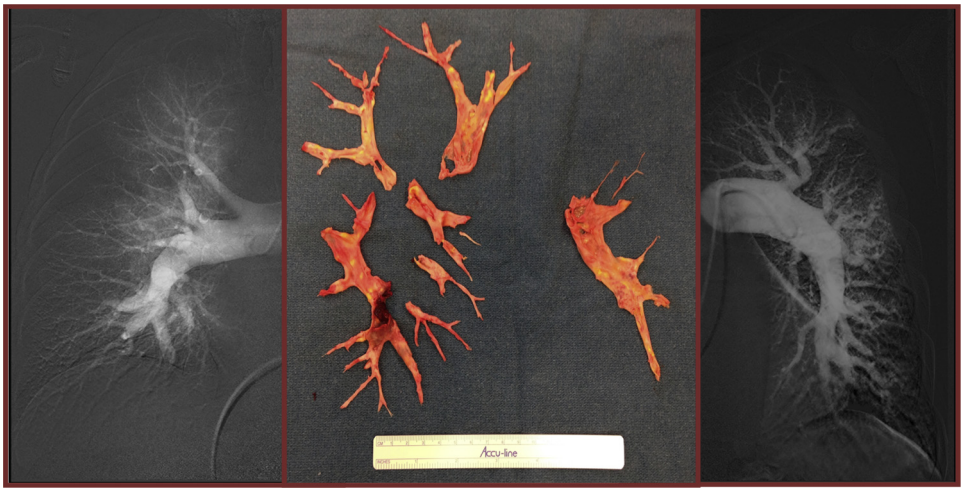

FIGURE 1. Pulmonary angiography tends to underestimate the amount of disease found at surgery in patients with CTEPH, particularly at the segmental and subsegmental level. Hence, in our experience, the correlation between the extent of thromboembolic disease and the severity of the pulmonary hypertension has been abandoned from the algorithm to determine surgical candidacy. The key factor associated with good outcome is a correct diagnosis and not necessarily the extent of disease on imaging.

(Figure 1). The key factor associated with good outcome is a correct diagnosis and not the extent of disease on imaging.

In conclusion, this paper presents important findings from the CHEST-1 study related to diagnosis and operability in patients with CTEPH. Accessibility to a second surgical opinion from experienced CTEPH centers is a crucial component in the evaluation of patients with CTEPH in clinical trials as well as in daily clinical practice, and patients with CTEPH should not missed this chance.

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