Editorial

Pulmonary thromboendarterectomy in chronic pulmonary disease—the Royal Papworth Hospital experience

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In this special edition of the journal, we review our understanding of chronic thromboembolic pulmonary hypertension (CTEPH) in 2021, and the progress made in the treatments over the last forty years. It has been an exciting time for developments in CTEPH, which has become the most treatable type of pulmonary hypertension with the availability of surgical, interventional and drug treatments.

Our own experience and some of the lessons we have learnt at Royal Papworth hospital are documented in the accompanying report (1). In retrospect, patients in the United Kingdom (UK) have been fortunate due to the foresight of the founding doctors and commissioners of our service. Two of the key principles were that the service was commissioned at a single centre for the whole of the UK to maximise experience, and that a patient follow-up protocol was built into the service specification so that the treating clinicians were able to learn from this experience. It is this direct feedback loop with evidence from detailed follow up that has allowed us to expand our indications for pulmonary thromboendarterectomy (PTE). As treatment options have widened for patients with CTEPH, the importance of a multidisciplinary team has increased. Within this team, the importance of expert radiology assessment is sometimes underestimated, but we feel that the importance cannot be overstated.

The above considerations are critically important when considering treatment for patients with chronic thromboembolic disease (CTED). These patients have symptoms of breathlessness and evidence of chronic thromboembolism on imaging, but they do not reach the threshold definition of PH at rest. Our group was the first to report our experience of PTE in this group of patients and coined the term (2). Three further centres have reported cases or case series since (3-6). However, since the 6th World Symposium in Pulmonary Hypertension, a new definition of PH has been proposed that has also changed the nomenclature for CTEPH and CTED. This new definition of PH as mean pulmonary artery pressure ≥20 mmHg with pulmonary vascular resistance of >3 Wood units (6) means that some of the patients included in the CTED case series above would now be classified as having PH. The proposed new nomenclature is that the disease is called chronic thromboembolic pulmonary disease (CTEPD), which includes both CTEPD without PH and CTEPH.

Irrespective of the name of the condition, the fundamental principle to understand is that PTE surgery in this patient group is only for symptomatic, rather than prognostic, benefit. If patients have no or mild PH at rest, there is no evidence at present for prognostic benefit from PTE surgery, but the potential complications remain as our report highlights. Although there is clinical experience of patients progressing from CTED to CTEPH with increasing PH within a few years, there is also evidence that many patients remain stable and may never develop PH. PTE is not a preventative treatment. Hence, the risk/benefit ratio for intervention is more difficult to calculate and the decision to operate is finely balanced with very detailed consent required. In our current clinical programme, we still offer PTE to these patients, but on a selected basis. We suggest that there should be a large burden of proximal disease, for example, occlusion on one side or involvement of both lower lobes, so that one can be relatively certain of

a good clearance of disease and a significant improvement in perfusion. Additionally, one should be confident that the patient's symptoms are explained by the thromboembolism and are not the result of other comorbidities. We would recommend a cardiopulmonary exercise test prior to considering surgery to ensure that exercise limitation is due to pulmonary vascular disease. The risk of the intervention should also be clearly defined and patients should not have additional risk factors for surgery. Thus, PTE for a patient without PH may be an excellent treatment for a younger athlete to restore quality of life but not appropriate in an elderly patient with arthritis and left heart disease, even if they have equivalent disease distribution on imaging. The lesson is that although these patients may appear to be the lowest risk for PTE surgery, they actually require more thought and careful assessment than those with established CTEPH.

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Footnote

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