## Diagnosis and Treatment of Chronic Thromboembolic Pulmonary Hypertension: An Era of Hope

Chronic thromboembolic pulmonary hypertension (CTEPH) may rank as one of the most underdiagnosed subsets among pulmonary hypertension categories. Designated as WHO Group 4 PH, CTEPH has been a notoriously difficult disease to determine from an epidemiological standpoint or to clearly elucidate mechanism and pathophysiology. CTEPH has also been shrouded in misconceptions and misunderstandings, such as whether a CT angiogram is sufficient to evaluate CTEPH; or the relationship of a likelihood of presenting with CTEPH; or that pulmonary thromboendarterectomy (PTE) evaluation applies to some selected patients with CTEPH. Rather, we now know that a VQ scan is the test of choice to "rule out" and diagnose CTEPH; a good portion of patients do not have a history of prior pulmonary embolism when they present with

## GUEST EDITOR'S MEMO

## Chronic Thromboembolic Pulmonary Hypertension: Current Care

This issue of Advances in Pulmonary Hypertension is dedicated to the topic of chronic thromboembolic pulmonary hypertension (CTEPH). It is imperative that all healthcare providers who care for patients with pulmonary hypertension be aware of the appropriate screening tests and treatment of this disease as CTEPH is potentially curable with surgery. Articles in this issue review the diagnostic evaluation, the surgical procedure of pulmonary thromboendarterectomy (PTE) (also referred to as pulmonary endarterectomy [PEA]), post-operative complications, care of surgical patients after hospital discharge, as well as the role of medical therapy of CTEPH.

Drs. Van Kan and Bresser provide an excellent and thorough review of the

CTEPH; and PTE should be considered for every patient who is diagnosed with CTEPH because it is the only form of PH that is potentially curable with a well-planned and appropriate surgical intervention.

It is thus my distinct pleasure to present this issue that focuses on the significant advances that have been made in diagnosing and treating this complex condition. I am very grateful to all the contributions of our Guest Editors, Dr. Richard Channick and Dr. Kim Kerr. They have assembled a renowned team of CTEPH experts to present the most current recommendations and emerging science focusing on diagnostic modalities, therapeutic considerations, and approaches-from specifics details of PTE surgery to discussing medical therapy with a focus on riociguat, which has been recently approved by FDA for treatment of patients with inoperable CTEPH or with recurrent PH post-PTE. The lively roundtable discussion, led by Dr Channick, who is joined by Drs. Auger, McLaughlin, Pepke-Zaba, and Tapson, covers some

diagnosis and preoperative evaluation of CTEPH. Perfusion scintigraphy is highly sensitive for CTEPH, and is therefore the recommended screening test for this form of pulmonary hypertension. However, perfusion scans are not specific for CTEPH and, therefore, any patient with pulmonary hypertension and an abnormal perfusion scan should undergo further imaging such as CT angiography, pulmonary angiography, or MR angiography to establish the diagnosis of CTEPH and the location/operability of the clots. Echocardiography is also used as a screening test for the presence of pulmonary hypertension, followed by right heart catheterization to better quantify the hemodynamic impairment as well as the potential benefits and risks of surgery.

Drs. Poch and Pretorius review the topics of operability assessment, the PTE surgical procedure, and surgical outof the controversial and difficult topics in treating patients with pulmonary thromboembolic disease such as thrombolysis and diagnostic dilemmas, to name a few. Dr. Studer provides a thoughtful commentary on the role of specialized care centers in the management of CTEPH, and Ms. Drcar gives us the nuts and bolts of caring for patients post PTE surgery.

Finally, we have the pleasure of introducing the Pulmonary Hypertension Care Centers (PHCC) initiative by Drs. Chakinala and McGoon, the first of a series of articles that discusses the fundamental rationale and workings of this accreditation process.

I hope you find this issue enjoyable and helpful in your care of this challenging group of patients.

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comes. Determining operability requires an assessment of not just the location of the chronic thrombembolic lesions, but a clinical decision on whether the thrombus burden correlates with the degree of hemodynamic impairment, as well as diagnosing comorbidities that might affect surgical outcomes. This article, as well as the preceding article by Drs. van Kan and Bresser, stress that operability assessment should only be performed at experienced centers. The surgical technique is described, allowing readers to appreciate why this surgery should only be performed by surgeons with expertise in PTE surgery. In experienced hands, this procedure results in sustained significant hemodynamic and functional improvement with an acceptable mortality risk. The most common post-operative complications