DIAGNOSTIC AND TREATMENT ADVANCES FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Alan B. Lumsden, M.D., Myung Park, M.D.

Houston Methodist DeBakey Heart & Vascular Center, Houston Methodist Hospital, Houston, Texas

Pulmonary embolism (PE) ranks after heart attack and stroke as the third most common cause of death from cardiovascular disease. Although anticoagulation therapy is effective in resolving most clots, a systematic literature review from 2014 estimated that approximately 4% of patients will go on to develop chronic thromboembolic pulmonary hypertension (CTEPH), a major complication of acute PE that carries extensive risk of morbidity and mortality. Preventing disease progression and morbidity associated with CTEPH hinges on early identification, proper patient selection, and meticulous treatment of these high-risk patients.

In this special issue of the *Methodist DeBakey Cardiovascular Journal*, we have compiled a compendium of information on the diagnosis and treatment of CTEPH provided by experts from the University of California, San Diego School of Medicine and our own institution. Because of the nonspecific nature of its two major symptoms, dyspnea on exertion and fatigue, CTEPH is severely underdiagnosed, and symptoms may not appear until the occurrence of right heart failure. Therefore, it is our goal to provide a general overview of the disease to broaden physicians' understanding, enhance their ability to determine a true diagnosis of CTEPH, and help them discern the most effective treatment based on selective patient criteria.

This issue opens with a review on the epidemiology and pathophysiology of CTEPH by Drs. Zeenat Safdar and Sarah Medrek. Excessive health care utilization, high-cost therapy, and increased mortality associated with CTEPH puts a significant burden on society, one that the authors hope to alleviate through enhanced awareness of this disease. By providing an overview of CTEPH origins, pathophysiology, and clinical risk factors, the authors aim to increase physician understanding of the disease process and help them to accurately and promptly identify and diagnose patients with CTEPH.

Early diagnosis is, in fact, paramount to increasing the likelihood of successful treatment. However, while CTEPH is the only form of pulmonary hypertension that is potentially curable with surgical or catheter-based intervention, it remains largely underdiagnosed. To address this conundrum, Drs. Humna Abid Memon and Ashrith Guha outline the parameters used for a comprehensive diagnostic workup, explaining how ventilation-perfusion scan, computed tomography pulmonary angiogram, and multiple other imaging modalities can be applied sequentially to render an accurate diagnosis. They also describe how concurrent use of right heart catheterization and pulmonary angiogram can determine operability and procedural risk for pulmonary endarterectomy as well as overall patient prognosis.

This issue then delves into the spectrum of treatments for CTEPH, starting with a review by Dr. William Auger and colleagues on the role of medical therapy and balloon pulmonary angioplasty. Although CTEPH is potentially curable with pulmonary thromboendarterectomy, almost a third of patients are determined to be inoperable—some due to technically inoperable chronic thromboembolic disease and others due to comorbidities that likely preclude a reasonable short- or long-term benefit. For these patients, both pulmonary hypertension (PH)-targeted medical therapy and balloon pulmonary angioplasty have demonstrated potential therapeutic efficacy. Both therapies have shown to improve hemodynamics, and some PH-targeted medical therapies have demonstrated improvement in 6-minute walk distance.

In terms of surgical treatment, minimally invasive surgery has revolutionized the entire spectrum of cardiac surgeries, from coronary bypass to aneurysm removal and valve replacement. It is no surprise, then, that it has been integrated into the therapeutic arsenal for pulmonary embolism as well. Two articles feature the range of surgical treatments available for patients with PE and CTEPH—one focusing specifically on pulmonary thromboendarterectomy and the other highlighting hybrid surgical and endovascular approaches. The first, by Dr. Michael Madani, describes the manifestation of CTEPH and explains the technique used in performing a pulmonary endarterectomy, a technically demanding yet potentially curative treatment for CTEPH that is available in only a few centers around the world. The review pays special attention to the factors that determine effective patient selection and reinforces how these, along with meticulous surgical technique and careful postoperative management, are the primary components contributing to the efficacy of this procedure. In the second, Drs. Alan Lumsden and Erik Suarez discuss currently available open surgical and catheter-based interventions and describe their own development of pulmonary embolectomy procedures to facilitate selective thrombus removal in the acute PE setting. They also introduce hybrid rooms that are used for both diagnosis and treatment of PE and explain how the creation of lightweight, easily deployable inferior vena cava filters used in conjunction with surgery has improved morbidity and mortality rates in patients with life-threatening pulmonary embolism.

As readers peruse this issue, they will notice a common consensus regarding the need for early diagnosis, proper patient selection, and substantial physician expertise to achieve favorable outcomes with CTEPH. It is our belief, and that of the contributing authors, that educating physicians to identify CTEPH and training them to master both patient selection and the techniques of pulmonary endarterectomy and other medical, surgical, and endovascular interventions should be a priority in the treatment of this disease.