Managing the Patient with Pulmonary Hypertension

Specialty Care Centers, Coordinated Care, and Patient Support

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KEYWORDS

• Pulmonary Hypertension Care Centers • Multidisciplinary care • Comanagement • Accreditation

KEY POINTS

- Pulmonary hypertension (PH) remains a challenging condition to detect, properly diagnose, and manage longitudinally in an era of ever-expanding therapies.
- The Pulmonary Hypertension Care Center program of the Pulmonary Hypertension Association was developed to address challenges in caring for patients with PH and improve the quality of care and long-term outcomes.
- Effective management of pulmonary arterial hypertension (PAH) requires close coordination between experienced practitioners from numerous disciplines at specialty referral centers and community-based practitioners.
- PAH patients' needs are best met by using the numerous health care resources that extend beyond the specialty care center.

INTRODUCTION

The field of pulmonary hypertension (PH) has evolved and expanded tremendously since the 1990s in conjunction with greater understanding of the condition, better appreciation of the epidemiology and, most important, the availability of numerous medical therapies specific for pulmonary arterial hypertension (PAH). In particular, PAH (or group 1 PH of the 5th World Symposium on Pulmonary Hypertension) has been transformed from a fatal disease with a very limited life expectancy to a chronic, manageable condition with a lifespan that can last years to decades.[1] As PAH patients live longer, unique management issues have arisen and large cohorts of patients have been compiled at specialty centers throughout the country. This article builds

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on many of the medical and scientific concepts that are relevant to this issue, by discussing evolving health care delivery for PH using a model of specialty centers staffed by multidisciplinary teams who strive to co-manage patients with local community-based physicians.

CURRENT CHALLENGES IN THE MANAGEMENT OF PULMONARY HYPERTENSION

Centralized Versus Decentralized Care

Historically in the United States (and still in many parts of the world), the care of PAH patients followed a centralized model with prompt referral of newly diagnosed patients to expert centers for comprehensive evaluation, accurate diagnosis, and access to advanced therapies, such as intravenous prostacyclin by home-based continuous infusion and lung transplantation. With greater awareness of PAH and development of many simpler pharmacologic options, management of PAH has shifted from tertiary referral centers to a broader range of health care providers and facilities thus leading to decentralized care. Concomitantly, PAH-specific therapies have been applied to increasingly diverse patients within group 1 PH and sometimes to non–group 1 PH patients, often leading to predictably poor treatment responses or even potential harm while increasing health care expenditures.

According to the Rare Disease Act of 2002 and the National Organization for Rare Disorders, any condition that affects fewer than 200,000 individuals at any given time in the United States constitutes a rare disease.[2] Multiple national registries (conducted outside of the United States) assert a prevalence of 15 to 26 cases of PAH per million inhabitants, thus making PAH a rare disease in the United States and the developed world.[3–5] However, World Health Organization diagnostic groups 2 and 3 PH are very common conditions encountered in the context of underlying cardiac or pulmonary disorders, such as heart failure, valvular heart disease, chronic obstructive pulmonary disease, interstitial disease, and obstructive sleep apnea. In fact, PAH represents a very uncommon form of PH that can be diagnosed only after a comprehensive evaluation to exclude other types of PH, especially the more prevalent group 2 and group 3 varieties.[6]

Challenges in Evaluating Pulmonary Hypertension and Diagnostic Errors

The evaluation of PH is a multistep and laborious process.[7] Challenges to completing thorough and accurate evaluations in patients suspected of having PH include the inability to perform some of the most critical diagnostic studies (eg, ventilation–perfusion scans and right heart catheterizations) as well as a lack of sufficient experience to analyze certain diagnostic studies, especially echocardiograms and the hemodynamic data from invasive right heart catheterization, both of which require a thorough understanding of the pertinent pathophysiology and adherence to strict protocols of measurement (Table 1).

Recent publications have shed light on some emerging challenges relative to overdiagnosis errors. In A Multi-Center Study Of The Referral Of Pulmonary Hypertension Patients To Tertiary Pulmonary Hypertension Centers (RePHerral) Study, conducted at 3 large university-based tertiary care referral centers in the United States, 98 of 140 referred patients had been assigned a definitive diagnosis of PAH before referral, but 32 (33%) were ultimately determined to be misdiagnosed. Fifty-nine patients had not undergone a prereferral right heart catheterization. Forty-two patients were started on PAH-specific medications before referral, but contrary to published guidelines in more than one-half of the instances.[8] The PAH Quality Enhancement Research Initiative project revealed underuse of guideline-mandated studies for the evaluation of PH, especially the ventilation–perfusion scan and right heart catheterization, which can greatly impact the ultimate PH diagnosis and treatment approach.[9]

Challenges with the Management of Pulmonary Arterial Hypertension

Medical treatment options have truly revolutionized the care of PAH patients. Presently, there are 13 medications from 4 different classes of therapies—endothelin receptor antagonists, phosphodiesterase 5 inhibitors, soluble guanylate cyclase stimulators, and prostacyclin modulators—approved by the United States Federal Drug Administration. Although all medications have been proven efficacious, numerous factors distinguish the agents, including route of administration, ease of drug delivery, side effect profile, and cost, leading to variable use of medications. Indeed, the simplicity of oral agents makes them the most commonly used, even though more complex parenteral therapies are quite effective and considered the preferred treatment for the most compromised individuals.[10,11] Furthermore, combination regimens with drugs from more than one class are increasingly being used based on the findings of recent large-scale studies.[10–15]