Pulmonary Hypertension Care Centers

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Our understanding and management of pulmonary arterial hypertension (PAH) has advanced tremendously over the last 30 years. Numerous scientific discoveries have helped to elucidate underlying mechanisms. Registries dating from the National Institutes of Health (NIH)-sponsored Patient Registry for Primary Pulmonary Hypertension (PPH Registry) of the 1980s to the recent REVEAL and French National registries have provided valuable information on PAH's epidemiology, natural history, risk factors, and prognostic indicators. Clinical classifications and diagnostic algorithms have been developed and periodically updated through international collaboration. Most importantly, numerous pulmonary vasomodulating drugs have been developed, and their widespread use has been associated with longer survival and improved quality of life. Nevertheless, a cure remains elusive.

Even though PAH is a rare disease with challenging therapies, the delivery of health care for PAH has been transformed. Historically, care followed a *centralized* model with newly diagnosed patients being expeditiously referred to expert centers for comprehensive evaluation, accurate diagnosis, and access to advanced therapies, such as epoprostenol and lung transplantation. With increased awareness of pulmonary hypertension (PH) and as pharmacologic therapies for PAH have become more accessible, the treatment paradigm has shifted from tertiary referral centers to a broad spectrum of medical practices. As a result, PH management is now delivered in a *decentralized* system by an increasing number of providers with varying degrees of expertise, leading to nonuniformity of care. Concomitantly, specific therapies have been applied to an increasingly diverse population of patients with PH. As a result, early access for patients to expert centers and assurances that optimal care is provided to all patients have become relevant concerns.

Recent publications have highlighted these emerging challenges. In the 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 subsequently determined to be misdiagnosed. Forty-two patients were started on PAH-specific medications prior to referral, and 24 of these therapies were contrary to published guidelines. Fifty-nine patients had not had a pre-referral right heart catheterization.[1] The PAH-QuERI project revealed underutilization of guideline-mandated studies for the evaluation of PH, especially the ventilation-perfusion scan and right heart catheterization.[2] Additional literature spotlights some shortcomings in the management of PAH patients. Evidence suggests that patients followed outside of a referral center (compared with the individuals already under the referral center's care) are treated with oral therapies longer, are more compromised and more likely to need urgent initiation of parenteral prostanoids, and have lower survival rates even after prostanoids are initiated.[3] This raises the question of whether reliance on oral therapies by nonexpert centers delayed the appropriate and timely use of parenteral prostanoids. Evidence from the REVEAL Registry demonstrates that a substantial number of patients in functional class III or IV within 6 months of death had not received parenteral prostanoid at the time of death, suggesting possible underutilization of the most potent and effective class of therapies.[4] Although these reports have shortcomings in terms of their small scale, retrospective design, or missing data, they appear to validate the perception of late recognition of PH, inaccurate diagnosis of PAH, untimely referral to expert centers, and inappropriate utilization of advanced therapies.

Two years ago, the Scientific Leadership Council (SLC) of the Pulmonary Hypertension Association (PHA) identified these emerging issues and advised its parent organization to develop and sponsor an accreditation program for PH centers in the US to harmonize and optimize management of PH. This course of action represented a shift in the PHA's approach. Historically, PHA focused on growing the PH community in an effort to enhance disease awareness and patients' access to care. But the evolving trends in health care delivery as described above have transformed PHA's perspective and provided the resolve to

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PH Care Centers
Providing excellence in pulmonary hypertension care

2-Center System
Criteria Categories

	PHA CCC Center of Comprehensive Care	PHA RCP Regional Clinical Program
Center Director	***	**
Center Coordinator	***	**
Program Staff & Support Services	***	*
Facility	***	*
Clinical Research	**	

*Indicates the number and/or rigorousness of criteria to be satisfied within a category

Figure 1

embark on this ambitious new course. A new initiative was spawned by the SLC's recommendations: one that would help address the many challenges facing the PH community. The initiative's mission statement is to *establish a program of accredited centers with expertise in pulmonary hypertension that aspires to improve overall quality of care and ultimately improve outcomes of patients with pulmonary hypertension, particularly pulmonary arterial hypertension, a rare and life-threatening disease*.

As a first step, the Pulmonary Hypertension Care Center (PHCC) Committee was formed and task forces were developed to: 1) develop criteria defining levels of expertise among centers, 2) explore funding, 3) formulate an implementation plan, and 4) design a patient registry. The PHCC Committee studied other disease-specific accreditation programs and benefitted greatly from understanding the organization and approach of the highly evolved and successful *Cystic Fibrosis Foundation (CFF) Accredited Care Centers*. Similar to CFF's program, the overarching objective of the PHCC is to improve the overall care of patients, which should translate into better long-term patient outcomes. Such a challenging yet laudable goal can be accomplished through several interlocking components:

- Increasing disease awareness
- Improving access to expert care
- Raising the level of care at ALL centers through increased adherence to published guidelines and consensus statements
- Providing a blueprint to prospective programs for becoming PH care centers
- Fostering collaboration among expert centers for managing individual patients and cultivating new research opportunities in the field
- Conducting center-specific and national quality improvement projects with the aid of a national patient registry

Unlike the CFF program, which began decades ago in a time of few CF experts and no specific therapy, the PHCC is developing in an era with many more practitioners having varying levels of expertise, practicing in diverse environments, and using a number of FDA-approved PAH-specific therapies. Clearly, PH is managed much more diffusely than CF still is. Therefore, any plan for accreditation has to recognize this existing heterogeneity, especially when access to expert care is so vital for patients, while still holding centers accountable to a set of standards acceptable to the majority of stakeholders.

To face this challenge, the PHCC Committee has approached its mission with a spirit of inclusivity, and has incorporated flexibility in the criteria and evaluation methods. As an example, the number of actively managed patients expected at a PHCC, which understandably is a crude manner of assessing experience and expertise, is specified in the criteria but will be interpreted in the context of mitigating factors, such as duration of the program's existence, regional population density, and proximity to other PH programs. In addition, the design for 2 types of centers (ie, Centers of Comprehensive Care [CCC] and Regional Clinical Programs [RCP]) is a central feature of the program that will hopefully maximize the eventual number of PHCCs across the country and enhance access to expert care. Both designations will be promoted

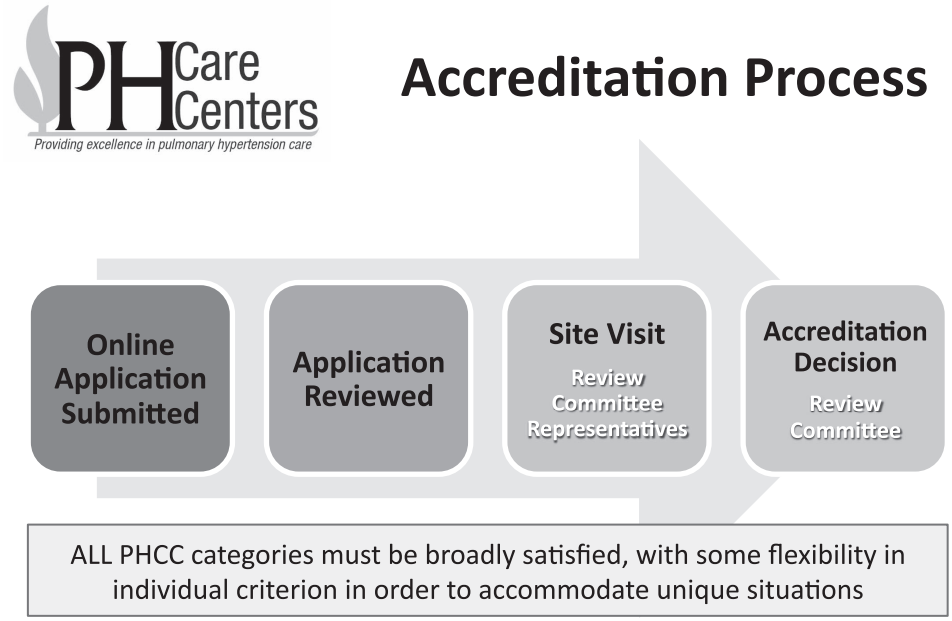

Figure 2

as PHA-accredited PHCCs and will have to meet their respective criteria through the same application process and evaluation method. Both types of centers will have to broadly satisfy several categories of criteria, including center director, center coordinator, program staff/support services, facilities, and research (CCC only) (see Figure 1 and visit www.phassociation.org/phcarecenters for more details).

Although inclusivity is a point of emphasis, the criteria and accreditation program recognizes the need for adherence to standards for selection of PHCCs, so that the designation represents a tangible achievement and conveys meaningful information to the relevant stakeholders. Achieving the optimal balance of *inclusivity* and *selectivity* has been challenging for the PHCC Committee and, understandably, no system can fully satisfy all interested individuals. Accordingly, it is important to note that the aggregate PH community will continue to be architects of the program and have the opportunity, through a well-developed governance structure, to modify and update the system as necessary. Clearly, the PHCC program will remain a work in progress for years to come.

Other obstacles for the PHCC program include securing reliable funding to initiate and maintain the program, as well as designing a reasonable rollout program to meet the perceived heavy initial demand for accreditation. From the outset, the PHCC Committee and the SLC mandated that the PHCC be free of pharmaceutical industry influence in order for the program to credibly maintain fairness and impartiality. Therefore, funding for this complex and intricate program will be derived from other sources, while not competing with other important PHA endeavors. It is anticipated that funding will rely heavily on accreditation fees from prospective and existing centers, similar to other disease-specific certification programs, and through fundraising efforts geared toward individuals and foundations. Moving forward, a sustainability committee will be assembled and will work with the PHCC governance structure to comprehensively procure new funding resources across the country.

The PHCC initiative already has generated tremendous interest in the United States, which is testimony to the perceived value of an accreditation program. By publicizing the criteria many months in advance, prospective centers will have an opportunity to enhance their respective programs. Significant demand and a large number of applications are anticipated once the program is inaugurated. In fact, an informal survey of PH Clinicians and Researchers (PHCR)/PH Professional Network (PHPN) membership in late 2013 revealed that *at least* 85 programs plan to apply for accreditation, with the majority hoping to apply in 2014. Accordingly, there is expectation of a flurry of applications to be received and site visits to be scheduled, with some unavoidable delay between application and accreditation (Figure 2). To minimize delays in the process and avoid unintended advantages to the "early" applicants, a sizable review committee is being formed and the program's accreditation announcements will likely occur in a batched manner that is still under discussion.

As the PHA and the PH community is on the cusp of launching this exciting and much needed grassroots program for accrediting PHCCs, it is vital to appreciate the enormity of the project and its potential consequences without becoming paralyzed by fear and uncertainty. For the sake of our patients, the PH community needs to find the courage and perseverance to forge ahead. In the next few issues of *Advances in Pulmonary Hypertension,* there will be additional updates about the PHCC program that will coincide with the PHCC's activities and milestones in the coming year.

References
1. Deano RC, Glassner-Kolmin C, Rubenfire M, et al. Referral of patients with pulmonary hypertension diagnoses to tertiary pulmonary hypertension centers: the multicenter RePHerral study. *JAMA Intern Med*. 2013;173(10):887-893.
2. McLaughlin VV, Langer A, Tan M, et al. Contemporary trends in the diagnosis and management of pulmonary arterial hypertension: an initiative to close the care gap. *Chest*. 2013;143(2):324-332.
3. Badagliacca R, Pezzuto B, Poscia R, et al. Prognostic factors in severe pulmonary hypertension patients who need parenteral prostanoid therapy: the impact of late referral. *J Heart Lung Transplant*. 2012;31(4):364-372.
4. Farber HW, Miller DP, Meltzer LA, McGoon MD. Treatment of patients with pulmonary arterial hypertension at the time of death or deterioration to functional class IV: insights from the REVEAL Registry. *J Heart Lung Transplant*. 2013;32(11):1114-1122.

GUEST EDITOR'S MEMO

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are hypoxemia and reperfusion lung injury, best managed by a multidisciplinary team.

The role of medical therapy for CTEPH is outlined in a very thorough review of the related literature by Drs. Rodriquez-Lopez and Channick. This is an important article given the observation that, despite lack of convincing data, the use of medical therapy for CTEPH prior to PTE has increased substantially over the past decade, culminating in the approval of a medication for patients with inoperable CTEPH or recurrent/persistent CTEPH following PTE.

This issue also includes a lively roundtable discussion by international CTEPH experts pondering many of the unanswered questions and debated issues surrounding CTEPH.

We hope you find this issue of Advances useful, and that it raises awareness and knowledge of this important disorder.

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