

Do Patients With Pulmonary Arterial Hypertension Benefit From Referral to a Specialized Center?

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Expert medical specialty care in the modern era took shape in the 19th century, according to an analysis by George Weisz,¹ as a result of clinical researchers seeking to “permit rigorous observation of many cases” of similar illnesses and to best manage illness at the population level. The development of medical specialties, and more recently subspecialties, provides a general endorsement of the concept that appropriate use of specialized care is beneficial. For a rare/orphan disease such as pulmonary arterial hypertension (PAH),^{2,3} the opportunity to improve care through specialized centers may be even greater since direct patient care experience outside such centers is uncommon. To justify the potential downsides including increased medical visit burden, traveling longer distance to the specialty center, and possible added costs, significant benefits of care at expert centers should be expected. To best support the argument for referral, this anticipated benefit should be evidence-based, include independent accreditation of specialty centers, and offer multiple opportunities to improve outcome and quality of life in PAH.

The fifth World Symposium on Pulmonary Hypertension (WSPH) recommends expert/specialized center referral with a 1-C grade, meaning that: “Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, or effective based on consensus of opinion of the experts

and/or small studies, retrospective studies, registries.”⁴ The authors based this recommendation on the observed success of the specialty center model for PAH care that has evolved in many countries,⁵ potentially improved access to possible emergency PAH treatments,⁶ and benefits of “high-volume specialized centers [that] have been recurrently shown to obtain the best outcomes for patients in different areas of medicine while maintaining greatest patient satisfaction, lowest complication rates, shortest length of hospital stay, and best value for health care payers.”⁷ The history of surgical therapy for chronic thromboembolic pulmonary hypertension (CTEPH) has also supported the concept of improved outcomes from high-volume centers: operative mortality has declined as centers gained greater experience through higher procedure volume.^{8,9}

Consistent with the WSPH recommendations regarding expert/specialty center referral, the Pulmonary Hypertension Association (PHA), through its Scientific Leadership Council, has developed a pioneering program, the PHA-Accredited Pulmonary Hypertension Care Centers (PHCC) initiative. This accreditation program has as its ultimate mission the improvement in outcomes for patients with PH. In addition, the PHCC program is intended to provide education that ensures best practice based on guidelines, to facilitate the establishment of care

registries, and to promote further clinical research. In their editorial published elsewhere in this issue,¹⁰ Chakinala and McGoon describe these efforts in detail from the PHCC Committee. These authors mention the impact of published study data such as that from the RePHerral Study, the PAH-QuERI project, and the REVEAL Registry, which suggest instances of diagnostic inaccuracy, underutilized guideline-based testing, and failure to prescribe parenteral prostacyclins ever or in a timely manner in patients with advanced disease. These gaps in PAH care present

Table 1. Potential advantages of referral to and collaboration with an expert specialty center for PAH

• Confirmation of diagnosis, especially in complex cases
• Initiation of treatment as early as possible
• Monitor therapy for adequacy of response
• Manage medication side effects
• Change or intensify therapy in a timely fashion
• Provide acute hospital care with access to all medications
• Evaluate RV function and stability over long term
• Improve connection to PHA and other support resources
• Offer participation by patients and families in PAH support groups
• Enhance awareness of and access to clinical trials
• Optimize access to and timing of lung transplantation

Abbreviations: PHA=Pulmonary Hypertension Association; RV=right ventricle

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opportunities for care obtained at accredited specialty centers to optimize PAH outcomes. Table 1 summarizes many of these anticipated benefits of referral to and collaboration with specialized treatment centers for PAH.

In summary, there is substantial evidence that referral for PAH care to specialty centers will improve outcomes. With the upcoming implementation of PHA accreditation of PHCC, patients and providers will soon have assurance that the designated specialty centers have the expertise they are seeking and deserve. This program will, I believe, reinforce the benefits of the medical specialty movement that began more than 150 years ago.

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