Pulmonary Hypertension Care Centers: Hope for the Future From a Patient's and Caregiver's Perspectives

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In 1987, after three years of being treated aggressively for asthma and then a prolapsed mitral valve, I was finally diagnosed with primary pulmonary hypertension (PPH). I was told I would be lucky if I lived 2 years without having a heart/double lung transplant and that I didn't have much time to get my affairs in order. There were fewer than 200 patients in the United States at the time. There were no treatment centers, support groups, or even an advocacy group like the Pulmonary Hypertension Association (PHA). I struggled with the loneliness of this diagnosis and the lack of information available.

I was 24 years old, very active, working full time in Connecticut, and taking some night classes. All of that came to what seemed like a screeching halt. I stopped school and focused my energy into contacting the National Organization for Rare Disorders (NORD) and National Institutes of Health (NIH) for information to give to my doctors, which, at that time, consisted of a transplant team. I attended support groups for transplant patients and waited to learn more about this illness and possible research opportunities. The information and research came many years later. Being a member of the PHA's Board of Trustees (BOT) and Pulmonary Hypertension Care Centers (PHCC) Oversight Committee is something I dared not dream of back then. –Diane

A few years earlier, across the country, Rachel was diagnosed with portal hypertension at age 10 following a massive esophageal hemorrhage. After battling back from this scare, my lovely sister began her journey with pulmonary hypertension (PH)—only no one knew she was on this journey. We, along with her medical providers, were unaware that portal hypertension could create another life-threatening condition.

Fast forward another decade or so: Rachel had experienced shortness of breath upon exertion for years, but she was repeatedly told by several physicians that she was out of shape and simply needed to exercise regularly. In August of 1993, a routine physical revealed a heart murmur and a "precautionary" EKG was done. At age 23, my sister was sassy, full of life, and unafraid. Thus, she went to her follow-up appointment alone, where she learned that although further tests were needed, she likely had something she had never heard of (inaccurately termed PPH) and that she should plan to make the most of the rest of her life—approximately 6 months. The physician who delivered this news had never seen a patient with PH, nor was there a single provider with knowledge of PH in the health system within which she was diagnosed. Rachel lived 18 months with her PH diagnosis before losing her battle in 1995, but clearly she had lived more than a decade with PH-related symptoms. –Laura

At the time that Diane and Rachel were diagnosed, there were no medical treatments for PH. Today, there are 12 FDA-approved treatments, with more on the way. Additionally, there is now a cadre of clinical and research experts who have devoted their careers to understanding PH and caring for those who are living with this complex illness. And, although there is evidence to suggest that many PH patients are living longer with a better quality of life, there are some key aspects of living with PH that have not changed for many patients.

For example, despite advances in understanding the etiology of pulmonary arterial hypertension (PAH), the time from onset of symptoms to recognition of the disease has not improved over the past 2 decades. Data from US adult patients enrolled in the Registry to Evaluate Early and Long-term PAH Disease Management (REVEAL) found that 1 in 5 patients (analytic sample n=2494 patients) with PAH experienced a delay of more than 2 years between symptom onset and either receiving a PAH diagnosis or starting PAH-specific therapy. This is particularly relevant given that untreated, PAH results in pathologic changes that are life-threatening and irreversible, and even a 2-year delay in diagnosis can greatly diminish the potential for good clinical outcomes and survival.[1]

The challenges associated with becoming diagnosed and accessing expert PH medical care are exacerbated for those living great distances from PH centers. These patients are often seen in community and nonspecialist settings for years before locating a PH expert either because they are limited in the ability to travel due to physical or economic constraints, or because their original treating provider is unaware of the complexities of PH and attempts to manage it without the involvement of a PH center. In some cases, patients are simply unaware of who and where the experts are. Additionally, the variability of the pathways leading to the different types of PH and the optimal treatments for

each of these make PH a highly complex condition to treat and manage successfully. Thus, those who are able to access centers with PH experience may encounter varied diagnostic approaches and treatment compared to similar patients across the country, or even in the next city or town.

For these reasons, the PHA BOT, through the recommendation and guidance of its Scientific Leadership Council, made the commitment to launch the PHCC initiative. This will ensure that a high standard of care is delivered at all PH centers, large or small, by reinforcing standards for practice as agreed upon by the medical experts in the field. This high standard of care is being assured through a PHCC Oversight Committee composed of expert PH clinicians and researchers, PHA BOT members, and patients. Additionally, a detailed center application process, accreditation standards and procedures, site visits conducted by volunteer reviewers with PH clinical and research expertise, and common data points for quality assurance evaluation are other ways in which high quality care will be achieved and monitored through this initiative.

From our unique perspectives, we believe that the PHCC initiative will: 1) promote diagnostic and treatment standards for PH; 2) reduce diagnosis and treatment errors, and improve overall quality of care; 3) increase PH-related knowledge among a broad array of medical professionals; 4) improve communication and linkage between centers; and 5) increase opportunities for additional research funding and activities.

What will this mean for patients and their families?

- Improved awareness of and access to centers with expertise in PH
- Increased information about available treatments
- Increased opportunities to participate in research
- More freedom to travel without the fear of having to see an inexperienced doctor in the case of an emergency
- Increased patient education
- Earlier and accurate diagnosis

In essence, having accredited PH centers across the country should eventually reduce the likelihood of delays in diagnosis, misdiagnosis, and insufficient or inappropriate treatment, and will ideally result in improved quality of life and longevity for many. It is our hope that the PHCC effort will ensure that future patients do not experience what Diane and Rachel did when beginning their journeys with PH.

Reference

1. Brown LM, Chen H, Halpern S, et al. Delay in recognition of pulmonary arterial hypertension: Factors identified from the REVEAL registry. *Chest* 2011;140:19-26.