


# Patient perspectives on pulmonary hypertension in the United States: Burdens, expectations, and goals

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## Abstract

Pulmonary hypertension (PH) is a life-threatening, debilitating disease caused by increased blood pressure in the pulmonary arteries. As patients living in the United States, we have unique insights into the journey from diagnosis and treatment within the US healthcare system and the significant impact that PH has on our quality of life. While there have recently been advances in PH management, there are several areas of PH care which we feel should be reassessed and improved. Commonly, diagnosis is lengthy and convoluted due to the rarity of the disease and limited knowledge of PH in primary care. There are also barriers to obtaining the right treatment and we feel that a more holistic approach to care is needed. Mental health is commonly overlooked and should be an integral part of patient care, as should elements such as nutritional advice, cardiopulmonary rehabilitation, and sexual health. PH patient associations play a key role in providing social, educational, and financial support to patients and caregivers alongside PH research and advocacy. As patients, we feel that we need to advocate for correct diagnosis, timely referral, and optimal treatment, in addition to overcoming the financial and/or administrative hurdles to obtain these. We propose several future goals to help empower patients to play an active, central role in their care and to improve all aspects of PH management. We advocate for further use of the patient voice in research and clinical development programs, including the use of patient-reported outcomes that have been developed with patient input.

## KEYWORDS

mental health, patient association, patient journey, patient-centered care, PH

**Abbreviations:** COPD, chronic obstructive pulmonary disease; CTEPH, chronic thromboembolic pulmonary hypertension; HCP, healthcare professional; HIV, human immunodeficiency virus; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; PHA, Pulmonary Hypertension Association; QoL, quality of life.

Kirsten Braley, Kathleen Richardson, and Lenise Whitley contributed equally to the manuscript.

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## INTRODUCTION

Pulmonary hypertension (PH) is a chronic, progressive disease characterized by elevated pulmonary arterial pressure at rest, which can result in debilitating symptoms, and ultimately right heart failure and death.<sup>1</sup> As patients with PH, we believe that understanding the patient's perspective of their PH journey is critical. It is important to provide the right information and support to patients and caregivers starting their PH journey, to empower patients to become their own care advocates, and to minimize barriers to diagnosis, referrals, treatment, and holistic care. Many of these aims are the focus of patient organizations, highlighting the critical role that they play in improving patient care. Understanding the patient perspective is also key to informing researchers, pharmaceutical companies, and healthcare professionals (HCPs) about gaps in current treatment and care pathways and how these may be rectified to improve patient care. A patient's perspective is also critical when developing or optimizing patient-reported outcome assessments in clinical trials, and many regulatory authorities now require inclusion of the patient voice early in regulatory decisions.

This review will discuss, from our perspective as patients with PH in the United States, what is most important to patients with PH regarding disease burden, diagnosis, treatment journey, and experience with insurers and payers. In doing so, we describe our experiences as patients and outline what we believe is the role of patient organizations, and our goals for the future in improving patient care.

## METHODS

The three patient authors (KB, KR, and LW) were identified by Team PHenomenal Hope as individuals with PH willing to be interviewed about their condition. The sponsor's US Advocacy Relations team then held an unstructured conference call with all five authors, who were asked to talk about their experiences with PH, their diagnosis, treatment journey, thoughts on patient care and unmet needs, experiences with patient associations, goals for PH in the future, and any other topics they found relevant. The authors from Team PHenomenal Hope (KW and LA) were also asked to talk about their experiences with patients and HCPs, and their goals for the future. Before the call, the authors were asked to provide a list of words that described them before and after their PH diagnosis, and word clouds were developed from their responses. After the call, the authors were free to contact the Advocacy Relations team with any

additional thoughts. The information from the authors was collated and literature searches were conducted on the points raised to gain the perspective of the wider community of patients with PH.

## Physical burden

PH causes a range of symptoms including dyspnea, fatigue, chest pain, weakness, weight loss, palpitations, and syncope.<sup>2,3</sup> In a European survey of patients with pulmonary arterial hypertension (PAH), 83% reported that they had difficulty climbing the stairs and 52% had difficulty walking short distances compared with before their diagnosis.<sup>4</sup> In the authors' experience, shortness of breath and a racing heart during normal activities such as walking, climbing stairs, or performing household chores is the most obvious physical burden of PH. Patients are forced to consider whether even short distances are walkable or whether they need to ensure that there is a suitable place to rest before starting to walk. Consequently, PH, regardless of etiology (e.g., PAH, left heart failure, chronic obstructive pulmonary disease, scleroderma, etc.), is associated with a substantial negative impact on quality of life (QoL).<sup>2,3</sup> The physical burden of PH impacts multiple aspects of a patient's daily life, including reduced physical capability to perform everyday tasks and hobbies.<sup>4,5</sup> In the PAH European survey, 83% of patients had difficulty with everyday domestic work and chores, with 72% finding that errands such as grocery shopping were impacted by their condition.<sup>4</sup> The physical burden of PH also impacts a patient's social activities; almost all patients (97%) taking part in the PAH European survey found that their ability to participate in sports and exercise was affected by their disease, with 77% and 55% reporting that vacations and leisure and cultural activities, respectively, were also affected.<sup>4</sup> In an international PH ethnographic study, patients were found to change their activities or hobbies to match their activity tolerance.<sup>5</sup> With appropriate medication, however, some patients do experience improvement in physical burden over time.

## Emotional burden

### Emotional impact of PH through the patient's disease journey

PH can significantly impact a patient's mental health throughout the course of the disease, but this is commonly overlooked as part of the required holistic care for PH.

While waiting for a diagnosis, patients commonly feel anger and frustration at the lengthy diagnosis or referral pathway. However, after diagnosis, their emotions change to feelings such as fear, confusion, loneliness, anxiety, and sadness due to the potentially life-threatening prognosis given and how PH may impact their and their families' lives (Figure 1). A key theme once diagnosed is the development of mental health conditions such as anxiety and depression, but there is a lack of awareness from HCPs in assessing and treating these conditions and a lack of mental health educational resources available to patients (Figure 2).

The mental health conditions experienced by the authors reflect those reported in studies assessing the impact of PH on mental health. In a literature review assessing the prevalence of anxiety and depression in patients with PAH, up to half of patients were reported to have depression (8%–53%) and anxiety or panic disorders (19%–51%), with the conclusion that mental disorders are commonly underestimated by HCPs.<sup>6</sup> Similarly, a PH study reported that 45% of patients had depressive symptoms, with common themes of psychological distress being “loss of myself,” “isolation from my surroundings,” “hassle associated with oxygen therapy,” and “fear of illness progression/deterioration.”<sup>7</sup>

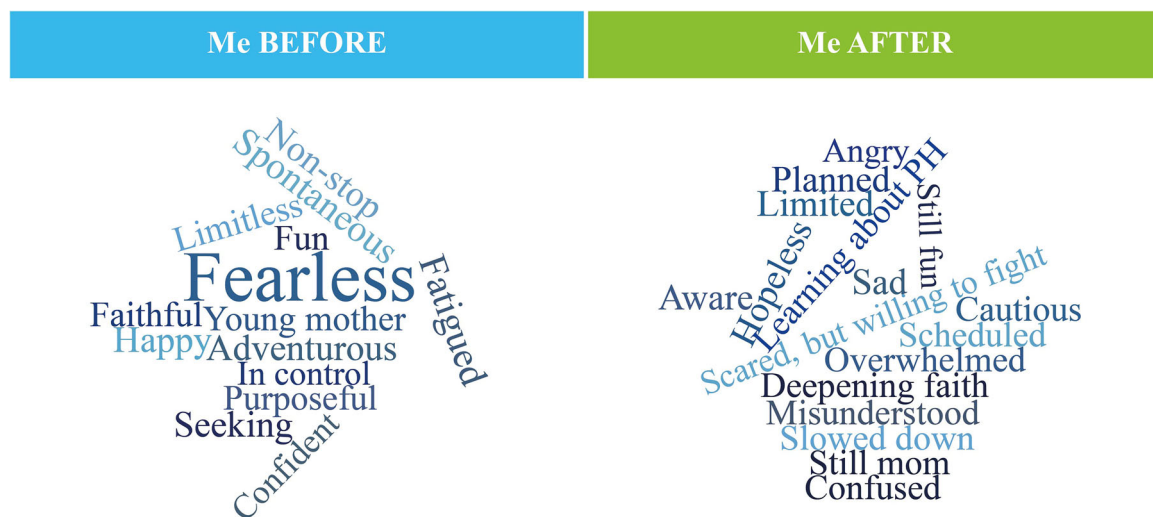
Other emotional aspects of a patient's life are negatively impacted by PH. Social interactions are affected, with a decreased ability to connect with friends and family and take part in leisure activities, as well as an increasing dependence on family members.<sup>4,5</sup> PH also impacts a patient's ability to work, adding financial burden to their emotional burden.<sup>4</sup>

Caregivers' social and emotional status is also negatively impacted, with common feelings of exhaustion, isolation, stress, and frustration.<sup>4,8</sup> PH may impact caregivers' ability to work, which can further exacerbate financial burden.<sup>4</sup>

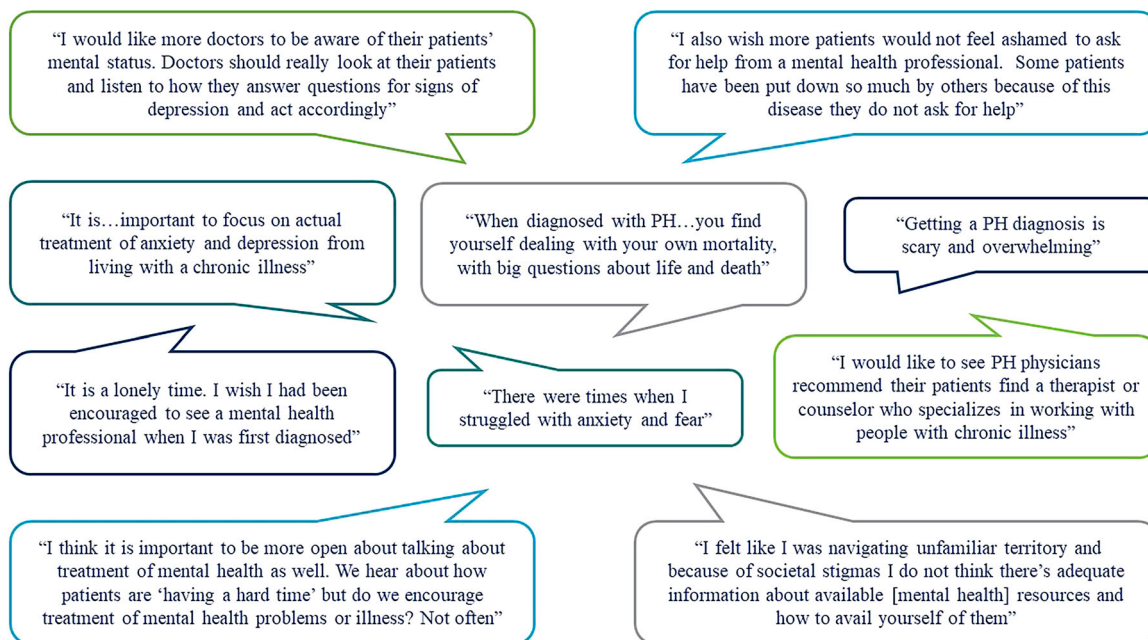
Sexual dysfunction and impaired sex-related QoL are often reported in women with PH, including a reduced frequency of sex after diagnosis of PAH and fear of sexual intercourse because of cardiopulmonary symptoms, as well as effects on relationships and the need to avoid becoming pregnant while receiving treatment.<sup>9–11</sup> Importantly, the impact of the disease on sexual health is often not discussed with HCPs and may not be captured by conventional QoL measures.

### Emotional impact of PH as an invisible disease

Many patients are told that they “don't look sick” (a commonly used phrase) and are not taken seriously by family members and friends. Consequently, patients may be labeled as attention-seeking, hypochondriac, or lazy because they cannot function in the way or time frame that they used to. Patients find themselves excluded from activities and gatherings because they are slower, always have to find a bathroom due to diuretics, or are sick and have to cancel planned activities. Feelings of isolation, depression, and anxiety are therefore common. These experiences are mirrored in published reports where patients with PH have reported that there is a lack of understanding and belief about the seriousness of their disease among their family and friends, leading to increased feelings of isolation, reported in 40% of



**FIGURE 1** Word cloud from the authors' responses to “What words describe ‘YOU’ before and after your PH diagnosis?” PH, pulmonary hypertension.



**FIGURE 2** Quotes from the authors about the impact of PH on mental health. PH, pulmonary hypertension.

patients, and a lack of empathy toward the patient.<sup>4</sup> Patients describe hiding their symptoms due to difficulties talking about their PH, the “invisible” nature of PH, and feelings of self-consciousness, fear, and hopelessness.

## Disease management burden

### Diagnosis

The diagnosis of PH for most patients is a lengthy process with a convoluted diagnostic pathway. In our collective experience, diagnosis took between 6 months and 2 years, although the longer time frame is more common. Published data support our experience of delayed diagnosis, with approximately a fifth of patients from the REVEAL Registry having had symptoms for >2 years before PAH was diagnosed.<sup>12</sup> A mean diagnostic delay of 2.5 years was reported for PAH from the Australian and New Zealand PH registry, with longer diagnostic delays being associated with decreased 5-year survival.<sup>13</sup> Similarly for chronic thromboembolic PH (CTEPH), a median diagnostic delay of 14 months was reported in an international CTEPH registry.<sup>14</sup> Patients often see several physicians before a diagnosis is reached, as evidenced by an Australian, single-center study that reported patients had on average approximately five primary care visits and three specialist reviews before being seen at a PH expert center.<sup>15</sup>

There are several reasons for this delay. Due to the rarity of PH, primary care and some specialist physicians

(e.g., cardiologists and pulmonologists) often lack an in-depth understanding of PH. When presenting to primary care physicians, patients are commonly not listened to or believed, particularly when tests were normal due to the disease being in its early stages. Physicians may assume that symptoms are anxiety or stress related, with the authors experiencing particular biases toward women. During a patient’s workup, implicit (unconscious) biases regarding age, body habitus, comorbidities, and gender may impact the diagnostic process. For example, it is not uncommon for patients presenting to physicians with shortness of breath to be told to lose weight. While obesity may certainly contribute to deconditioning, its presence should not delay a search for alternative causes of symptoms. Supporting our observations, data from the REVEAL Registry show that age and comorbidities were risk factors for delaying diagnosis of PH.<sup>12</sup> Gender, race, age, socioeconomic status, and weight biases are also commonly prohibitive to reaching a correct diagnosis in several other disease states.<sup>16–19</sup> Implicit biases among physicians can influence the quality of care given to patients; for example, studies have shown that Black and Hispanic patients receive less pain relief in the emergency department than White and non-Hispanic patients, respectively.<sup>20</sup> Removal of these misconceptions and assessment of each patient as an individual along with their specific symptoms and consideration of PH in patients with comorbidities should be aspired to.

Another reason for delayed diagnosis is that PH is commonly misdiagnosed, likely due to the nonspecific symptoms (e.g., shortness of breath) associated with

early-stage disease.<sup>21</sup> An author in our group was diagnosed with anemia and then 2 months later asthma (a common misdiagnosis), before 2 months later being given a final diagnosis of PAH following a hospital admission. In line with our experiences, studies have shown that a third of patients with PH receive a misdiagnosis, and between half and almost two-thirds are classified with more severe disease (World Health Organization functional class III/IV) at diagnosis, suggesting delayed diagnosis, prior misdiagnosis, or late referrals for these patients.<sup>22–24</sup>

Given these issues with delayed and/or misdiagnosis, patients must commonly advocate for their own diagnosis and encourage physicians to send them for further tests and consider different diagnoses.

## Referral and treatment

Once diagnosed, patients must frequently advocate for their own care so as to obtain the right diagnosis and treatment—a process that can be time-consuming and emotionally exhausting. Patients commonly feel they are not listened to by primary care or specialist physicians, who may not have the necessary knowledge or experience to provide the best care in terms of referral and treatment. Importantly, not all pulmonologists and cardiologists are PH specialists. A key part of the PH treatment journey, therefore, is finding a PH specialist; however, it is common for patients to have to proactively research (e.g., through family/friends or online) to find a PH specialist for referral, either in line with or against the advice of their care team. Importantly, patients may not receive the most appropriate treatment for their condition until they have seen a specialist. In an international survey of patients with PAH, 47% of patients said that they wanted information about specialists and doctors involved in PAH care.<sup>4</sup> Unfortunately, patients have also reported being prevented from getting a referral to an expert center due to insurance limitations.

Patients have reported being diagnosed with PH but that the physician did not follow up with them about the diagnosis, as they thought that there was no treatment or that treatment was not needed as the disease was mild. Published data also support our experience that patients with PH do not always receive the recommended treatment. In a study of patients referred to specialist PH centers, almost a third started receiving a PAH therapy before referral, and of these just over half had been prescribed therapies contrary to treatment guidelines.<sup>22</sup> In an international CTEPH physician survey, a high proportion of patients with CTEPH were reported to be using off-

label drugs, with 17%–55%, 19%–56%, and 52%–86% of patients prescribed a prostanoid, endothelin receptor antagonist, and phosphodiesterase 5 inhibitor, respectively, depending on the region.<sup>24</sup> Furthermore, in almost half of patients, the curative, recommended, and approved surgical option, pulmonary endarterectomy, was only considered an option when drug therapy had failed, contrary to treatment guideline recommendations.<sup>1</sup>

Oral medication regimens can be complex, with patients often requiring multiple drugs to manage their PH.<sup>1</sup> A patient's concerns about potential adverse effects of PH treatments can significantly impact adherence.<sup>25</sup> Indeed, a recent meta-analysis of 14 studies in patients with PAH found that 39% of patients were not fully adherent to their medication regimen, with 42% of patients discontinuing medication.<sup>26</sup> Parenteral medications can also add to the patient's burden as they can require patients to carry pumps with them, manage pump settings, and maintain the equipment and site of administration in a sterile manner,<sup>27</sup> which can impact daily activities and increase the visibility of a patient's disease.<sup>28</sup> Such routes of administration are also associated with side effects such as injection site pain, catheter-related sepsis, and thrombosis.<sup>29–32</sup> Patients have reported being concerned regarding the speed of intravenous medications reaching the circulatory system and any potential deleterious effects associated with this. Collectively, these issues can lead to poor treatment adherence and negatively affect QoL. The need for supplemental oxygen also adds to the patient's burden; for example, additional planning is needed when leaving the house to factor in taking an oxygen tank, including calculating the duration a patient will be away from home to ensure they have taken enough tanks with them.

As PH progresses to severe right heart failure, advanced treatment options are needed to support these patients. Such palliative care includes invasive interventions such as atrial septostomy, noninvasive therapy such as symptomatic management of pain and respiratory distress, and emotional and spiritual care with a pastoral counselor,<sup>33</sup> all of which are important for providing the best possible QoL at end of life. However, in a survey of PAH physicians' attitude toward palliative care, awareness and utilization of palliative care were low, with misconceptions about the scope and goals of palliative care.<sup>34</sup> In providing these complex treatments at the right time for the right patients, difficult conversations are needed between the physician and the patient, and their family and/or caregivers. However, physicians typically receive very little training in handling these difficult situations. Further guidance is therefore needed to support physicians and other HCPs in effectively communicating the challenges and reality of these

treatments and how best to transition to the patient's goals of care.<sup>35</sup> There is also a lack of adequate hospice care at end of life given the high cost of PH medications that are not covered under most hospice programs. These problems represent a large gap in the care and management of patients with PH.

## Treatment costs and insurance

Depending on where they live or receive care, many patients will have to interact with medical insurance companies to facilitate their care. Insurance pre-authorizations are required for many PH treatments, which newly diagnosed patients may not be aware of. These can be challenging to secure, particularly if insurance providers are changed. For authorization, cumbersome documentation must be submitted by physicians showing diagnostic and test results justifying the need for the PH medication; this documentation must be updated annually or whenever physicians adjust the dosing. Missing information can lead to delays in the pharmacy filling the prescription, and pharmacies are not always proactive in asking for missing information. As with diagnosis and treatment, patients must be proactive with their physicians and pharmacies to navigate the hurdles with insurance companies to cover prescriptions and avoid delays in refills.

Many patients with PH must also ensure they have a supply of supplemental oxygen. The high volume of paperwork associated with securing oxygen therapy can take time and energy, and result in delays for the patient. Patients commonly experience difficulties with oxygen supply companies in obtaining a regular and sufficient supply. Unfortunately, oxygen supply companies commonly have very limited customer service, with no systems in place to support patients directly. Furthermore, depending on location and insurance coverage, patients may have a very limited choice of oxygen suppliers, with no option of choosing a company that provides better support. Currently, there are also additional challenges in obtaining oxygen therapy due to the Covid-19 pandemic.

There are health disparities across different demographics of people affected by PH, which can be driven by financial and/or insurance factors. A PH study reported that Black patients were less likely to have private insurance versus Medicaid and had worse right ventricular function at diagnosis compared with White patients. Black race was associated with twofold increased mortality compared with White race after adjustment for age and functional class; however, this was not the case after adjustment for insurance status,

suggesting that insurance status can play an important role in PH outcomes.<sup>36</sup>

The challenges patients face in coordinating between drug companies, insurance companies, and HCPs add further burden and further exacerbate anxiety and depression. For patients who are unable to negotiate with insurance companies, disease progression may be the inevitable outcome. In the United States, PH is associated with a high financial burden with high out-of-pocket costs. Unfortunately, in most states, co-pay assistance in the form of grants from charitable foundations or coupons from drug companies are not allowed to count towards a patient's annual deductible or out-of-pocket cost-sharing limit, thus leaving patients still unable to afford care. Many patients rely on co-pay assistance from drug manufacturers or assistance groups, but this assistance is not always guaranteed.

## Currently available patient resources

At the sixth World Symposium on PH in 2018, a new task force on the patient's perspective was created that called for an expansion of patient associations to support patients and caregivers, lobbying for access to the best care and treatments, and input into the development of clinical trials and registries.<sup>37</sup>

PH patient associations can play several roles to support patients individually and collectively by driving improvements in PH care and management. These include patient and caregiver/family education, social support, financial support, research, and advocacy (Table 1). Other PH associations are active in other countries; of note, the Pulmonary Hypertension Association (PHA) Europe has been active in lobbying the European Parliament to raise awareness of the need for policy change and resources to improve PH management and the QoL of patients.<sup>38</sup>

Patient associations are often key in a patient's journey, providing trustworthy information to enable them to learn about PH and available treatments and care, particularly as patients commonly need to advocate for their own health care. Published data also demonstrate the importance of PH associations for caregivers, with approximately two-thirds of caregivers joining a patient association and a quarter reporting being active members. Just over half of those who joined a patient association did so to receive PAH education.<sup>4</sup>

Patient associations such as Team PHenomenal Hope and the PHA also provide social support through direct contact with other patients, caregivers, and support groups, and access to other patients' stories. Individual institutions may also offer support groups, such as the

**TABLE 1** Summary of US PH patient association resources and programs.

PH association	Mission/goals	Resources	Programs/events
Team PHe-nomenal Hope	<ul style="list-style-type: none"> <li>• Provide a novel platform of endurance athlete and patient partnerships in order to amplify the stories of PH</li> <li>• Provide seed funding and support to grassroots chapters around the world who share our commitment to raising awareness of PH and improving both patient care and QoL</li> <li>• Develop sustainable funding programs that support medical research and innovative means of patient support</li> </ul>	<ul style="list-style-type: none"> <li>• Patient stories</li> <li>• “What is PH?”: information on symptoms, diagnosis, classifications, and treatments</li> <li>• Online support group meetings with informative presentations from nurses and physicians</li> </ul>	<ul style="list-style-type: none"> <li>• Let Me Be Your Lungs (athletes are matched with volunteer patients to raise awareness about PH and the need for improved treatments)</li> <li>• Unmet Needs Patient Impact Fund (direct-to-patient grant to support nonmedical immediate hardship needs)</li> <li>• PHe-nomenal Impact Fund for Global PH Research (supports grants for medical research into improving treatments or finding a cure for PH)</li> <li>• International Team Development (international rollout of Let Me Be Your Lungs)</li> <li>• PH Feels Like awareness campaign (understanding PH from the patient’s perspective)</li> </ul>
Pulmonary Hypertension Association	<ul style="list-style-type: none"> <li>• Mission: to extend and improve the lives of those affected by PH</li> <li>• Vision: a world without PH, empowered by hope</li> </ul>	<ul style="list-style-type: none"> <li>• PHA Classroom (online education platform covering medical and lifestyle topics for patients and their families)</li> <li>• Educational resources (disease background, diagnosis, treatment options)</li> <li>• Tips for coping and living with PH</li> <li>• Patient stories</li> <li>• Research and clinical trial information</li> <li>• Insurance guide</li> <li>• Information on physicians and PH care centers</li> <li>• Information tailored to support patients, caregivers, and parents</li> </ul>	<ul style="list-style-type: none"> <li>• Support groups</li> <li>• Phone and online support</li> <li>• PHA Live (monthly series of Facebook Live events and webinars with PH expert speakers)</li> <li>• Advocacy Action Center</li> <li>• PHA research programs (Aldrighetti Research Award)</li> <li>• PHPN Symposium</li> <li>• PHA Registry</li> <li>• PHA Online University (CME-accredited e-learning website for healthcare providers)</li> <li>• PHA Medical Education On-Demand Program</li> <li>• PHA Preceptorship Program</li> <li>• Heart2CurePH awareness campaign</li> </ul>

Abbreviations: CME, continuing medical education; PH, pulmonary hypertension; PHA, Pulmonary Hypertension Association; PHPN, PH Professional Network; QoL, quality of life.

University of Iowa, Ohio State University, and Loma Linda University. Of note is the Let Me Be Your Lungs program from Team PHe-nomenal Hope, where athletes are matched with volunteer patients to raise awareness of PH, the impact of living with PH, and the need for improved treatments. Social support is also available from PHA support groups, as well as a telephone support line and an email initiative connecting members of the PH community (Email PHriends). Patients’ need for social support from patient associations is evidenced in an international survey of patients with PAH, where 32% of patients reported wanting testimonial and patient stories and 28% wanted patient association contacts.<sup>4</sup>

Furthermore, in an international ethnographic study, patients who had a connection with their national PH association reported feeling less isolated.<sup>5</sup> Social support for caregivers is also reported to be low, especially for emotional support, which was associated with more severe depressive symptoms in caregivers.<sup>39</sup> In an international survey of caregivers for patients with PAH, half of caregivers who joined a patient association did so to meet other patients and caregivers to understand how they were coping.<sup>4</sup>

Patient associations also offer information on PH care centers, PH expert centers, specialist HCPs, and specialist advice on financial assistance and coordinating with

insurance providers, pharmaceutical companies, and oxygen suppliers. For example, the PHA runs a monthly series of Facebook Live events and webinars with PH experts, as well as providing general educational resources on their website. Patient associations can directly offer financial support to the patient community in the short and long term. This includes patient grants, such as Team PHe-nomenal Hope's Unmet Needs Patient Impact Fund, which is a direct-to-patient hardship grant to help the critical nonmedical needs of patients with PH. Similarly, organizations such as the Patient Access Network Foundation provide financial assistance for patients with rare diseases, including PH. For more long-term projects, research grants and programs are available, such as the annual PHe-nomenal Impact Fund for Global PH Research and the PHA Aldrighetti Research Award for young investigators, which support grants for medical research into improving treatments or finding a cure for PH.

In addition to funding research, patient associations are actively involved in other aspects of PH research. These include or have included providing information on and participation in the development of PH-specific patient-reported outcomes. For example, in the development of the emPHasis-10 questionnaire, patient recruitment included those identified from the database of the PHA UK.<sup>40</sup> They also provide information on clinical trials and participation in clinical trials, and in the case of the PHA run a registry. The PHA Registry is the largest active US PH patient registry collecting diagnosis, clinical status, and treatment data from patients with PH who are starting evaluation and/or treatment.<sup>41</sup>

PH patient associations, in partnership with patient advocates, can also play a key role in raising awareness of PH as there are no more powerful advocates than patients themselves. Patient associations can support and empower patients with the knowledge to help drive change and enable better health outcomes and equity. The PHA provides information to understand the legislative process and encourage patients to advocate for healthcare policies that, for example, secure or improve access to treatments and help or limit healthcare costs for patients with PH.<sup>42</sup>

## Goals for the future

Although many advances have been made in terms of new treatment options and increased understanding of PH, we advocate for further improvements in patient management and care from the patient's perspective (Table 2). We discuss below several goals for the future from our perspective of living with PH.

## Disease awareness for HCPs

More education is needed, firstly, for physicians, both at the primary care and subspecialty level to allow for earlier diagnosis of PH, so that at presentation the right questions are asked to consider PH diagnosis without bias and how symptoms can masquerade as other diseases. Physicians should play an active listening role in patients' stories, so that the patient voice is truly heard through their own unique physical, psychological, and psychosocial experiences of PH, to facilitate faster diagnosis and referral. A set of learnings for PH diagnosis from our perspective is presented in Figure 3.

In addition to physicians, it is important not to overlook the education of other HCPs in the management of PH. Advanced-practice providers, such as nurse practitioners and physician assistants, as well as therapists and social workers, play key roles in the management of PH,<sup>43</sup> and thus educational programs should not focus exclusively on physicians. The need for holistic care in PH is discussed in more detail below.

## Patient empowerment

A key theme from patient experiences is the need to advocate for their own diagnosis and disease management, while navigating the complexities of referrals and being prescribed and obtaining the correct treatment. Ideally, patients should have an active collaboration with their physician and other HCPs, with the patient consulted on what their goals are and proactively informed of new treatment options. Importantly, for this collaboration to be successful, patients should be educated by their HCPs and/or patient associations so they can understand the information provided, be their own advocates, and participate in the formulation of their personalized treatment plan. Providing information at the time of diagnosis is key to helping alleviate anxiety and empower patients, as demonstrated in an international survey of patients with PAH, where 83%, 80%, and 77% of patients reported that diagnosis was the most suitable time to receive information about the disease, treatment options, and specialists involved in PAH care, respectively.<sup>4</sup>

Education for caregivers or family members is also essential to help them support the patient and to understand PH and the effect it will have on their lives.

## Mental health support

Mental health conditions are a common occurrence in patients with PH, but they are frequently underestimated



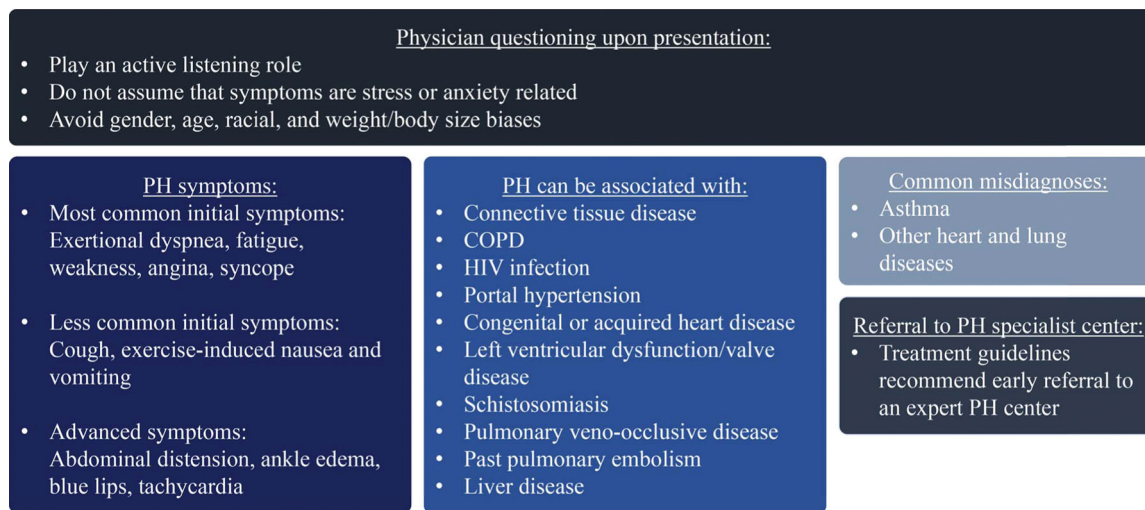
**TABLE 2** Key goals for improving the management and care of patients with PH.

Future goal	Recommendations
Increased disease awareness to reduce misdiagnosis and time to diagnosis, and to help patients and caregivers/families to understand PH and how to cope with it in everyday life	<ul style="list-style-type: none"> <li>Put posters in the offices of primary care physicians (and cardiologists) about PH and its red flags</li> <li>Educate primary care physicians about common biases or preconceptions during diagnosis</li> </ul>
Empowering the patient to be their own advocate	<ul style="list-style-type: none"> <li>Provide motivational information to patients or their dedicated caregivers to encourage them to take control of their own health care</li> <li>Provide patients and caregivers with educational materials about PH (the disease, treatments, expert centers) and how to connect with PH associations for further support (social, emotional, financial)</li> </ul>
Increased mental health awareness, help, and funding	<ul style="list-style-type: none"> <li>Train PH healthcare professionals to ask patients about their mental health</li> <li>At diagnosis, provide resources and advice to patients about how to choose a mental health professional</li> <li>Provide access to mental health help resources including educational and practical information, tools, tips, mental health support groups, and a hotline run by a social worker or counselor familiar with grief and chronic illness counseling</li> </ul>
Emotional support for caregivers	<ul style="list-style-type: none"> <li>Train or highlight the need for PH healthcare professionals to ask caregivers about the impact of PH on their lives and any support that they may need</li> </ul>
Holistic patient care	<ul style="list-style-type: none"> <li>Provide comprehensive patient care including access to mental health support, nutritional counseling, and cardiopulmonary rehabilitation within every PH clinic</li> <li>Further research to understand the nutritional and cardiopulmonary rehabilitation needs of patients with PH and the outcomes from these interventions so appropriate advice can be given to patients</li> </ul>
Financial support	<ul style="list-style-type: none"> <li>Education for payers and policymakers to allow patients to access medications more easily and affordably</li> </ul>
Involvement of the patient voice in PH research and clinical development	<ul style="list-style-type: none"> <li>Provide patients with information about research or advocacy programs and how to get involved</li> </ul>
Improved patient support from oxygen supply companies	<ul style="list-style-type: none"> <li>Improve customer service and patient support</li> <li>Provide timely and adequate responses to patient queries and complaints</li> <li>Commit to delivering oxygen supplies when agreed, ideally at a specified time, so that patients do not run out of oxygen and are not confined to their home waiting for a delivery</li> <li>Be prepared to discuss a patient's needs with the prescriber; do not rely only on Medicare guidelines to determine eligibility for oxygen</li> </ul>
Improved Medicare oxygen therapy guidelines	<ul style="list-style-type: none"> <li>Medicare guidelines for oxygen therapy should be updated so they are applicable to all patients with PH</li> </ul>

Abbreviation: PH, pulmonary hypertension.

and underdiagnosed by treating physicians, and limited information regarding mental health resources is available to patients. Further investment, resources, help, and awareness in PH mental health are therefore needed for patients and caregivers. Valuable mental health help resources would include educational and practical

information, access to counseling, therapy, and mental health support groups. We also suggest that patient care should include routine psychological screening and intervention, in accordance with the European Respiratory Society/European Society of Cardiology guidelines for PH management.<sup>1</sup> HCPs should be encouraged to ask



**FIGURE 3** A summary of important information on PH diagnosis for primary healthcare professionals.<sup>1</sup> COPD, chronic obstructive pulmonary disease; HIV, human immunodeficiency virus; PH, pulmonary hypertension.

open questions to patients to enable open discussion of mental health without stigma. This in turn should provide patients with easier access to treatment for anxiety and depression. HCPs should recommend therapists or counselors who specialize in working with chronic illness so that mental health counseling and treatment become an integral part of the patient care team. We would also advocate for a support hotline to be made available to patients as soon as they are diagnosed, providing access to a social worker or counselor familiar with grief and chronic illness counseling.

Emotional support is also needed for caregivers, and to this end HCPs should also be trained to ask caregivers about their mental health needs and any support that they require.

## Holistic care

To optimize patient outcomes, a more holistic approach addressing all aspects of a patient's life is recommended. In addition to the usual specialist, multidisciplinary medical team, other aspects of care such as mental health, sexual health, nutritional counseling, cardiopulmonary rehabilitation, and palliative care should be included in the care team, with the patient being a central member, empowering shared decision-making (Figure 4). Ideally these services should all be available within the PH clinic for easy access for the patient.

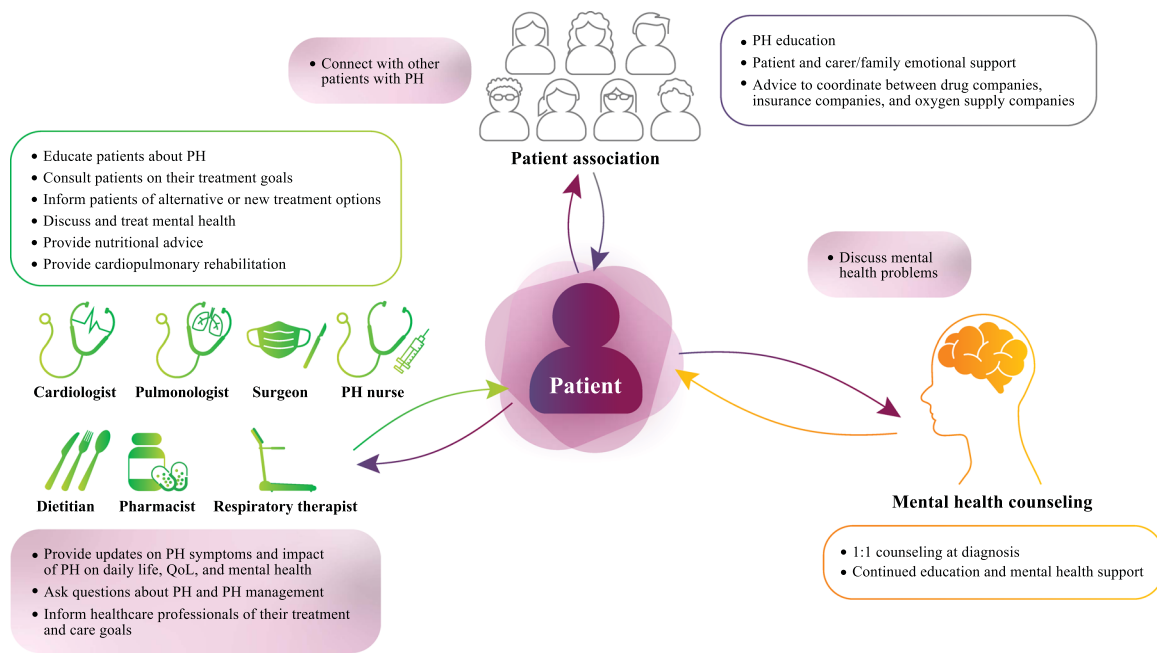
The use of cardiopulmonary rehabilitation in PH may be controversial for some physicians, but as long as exercise is tailored to individual patients and they understand how to exercise safely, we believe that all patients with PH can exercise and benefit from exercise. Cardiopulmonary

rehabilitation can also be integral in helping to determine supplemental oxygen needs for various types of exertion. Indeed, published data have suggested that exercise interventions can be effective in patients with various forms of PH but must be performed in a closely supervised setting.<sup>44,45</sup> The European Respiratory Society/European Society of Cardiology guidelines for PH management also recommend that supervised exercise training should be considered in physically deconditioned patients with PAH under medical therapy.<sup>1,6</sup> However, in a UK, survey-based study, while patients with PH and physiotherapists noted the benefits of physical activity, cardiopulmonary rehabilitation services were reported to be only available in acute inpatient care facilities, and specialist knowledge of PH rehabilitation/physiotherapy was limited in community services.<sup>46</sup>

Regarding the role of nutritional status and advice, limited evidence suggests that some patients with PAH have nutritional deficiencies, and changes to correct these may reduce disease burden.<sup>44,47</sup> Vitamin D and iron deficiencies are common in patients with PAH and have been shown to correlate with disease severity and prognosis.<sup>47,48</sup> Further research is needed to understand the nutritional needs of patients with PH so that appropriate advice can be given to patients. As there are specific body mass index guidelines for some PH treatments, such as lung transplant, nutritional advice should be given at diagnosis.

## Financial support

PH can result in financial burden for patients and caregivers in terms of treatment costs and their inability to work. Education for payers and policymakers is



**FIGURE 4** Schematic illustrating the “right” patient-centered care team with all specialties (including patient associations) needed for optimal care and bidirectional flow of goals/information between the patient and healthcare professionals. PH, pulmonary hypertension; QoL, quality of life.

therefore needed to allow patients to access medications more easily and affordably.

## PH research and clinical development

Further research and clinical development programs are key to advancing PH management, and the patient voice is becoming an important guide for pharmaceutical companies to know where to invest and how to guide research. The Food and Drug Administration is embracing patient-reported outcome assessments in clinical trials, and regulators now require that there is an early interaction with patients to develop tools that assess the impact of disease on QoL and the effect of treatments on symptoms.<sup>28,49,50</sup> Another future goal should therefore include involvement of the patient voice in PH research, with pharmaceutical companies designing clinical trials that access patients’ experience and QoL in response to treatments. Further research is also needed in patients with World Health Organization groups 2, 3, and 5 PH, for whom there are currently no approved treatments. Research addressing patient adherence issues with some treatments (e.g., improved oxygen tank design to better suit the everyday needs and lifestyles of patients) would also be beneficial.

Finally, we feel that researchers, pharmaceutical companies, and HCPs should try to empathize more with the people they are responsible for treating.

To achieve this goal, industry partners could be more involved in working with patient associations on projects together, such as matching with a patient with PH in the Let Me Be Your Lungs program. Such initiatives would provide more awareness of what it is like to live with PH.

## CONCLUSIONS

Improvements in diagnosis are needed to avoid delays and multiple, unnecessary tests, which could be accomplished with further education for primary care physicians. A common theme from patients is the need to advocate for their own care, from seeking a correct diagnosis through to all medical and financial aspects of treatment. To have access to the best care available, further support, in terms of education and HCP interactions, is needed to empower patients to become their own advocates. PH impacts many aspects of a patient’s life, therefore a holistic approach to treatment with a multifaceted care team should be the ideal standard. Mental health is commonly overlooked in PH and should be actively incorporated into this care team. PH patient associations are an important resource for patients as they play a key role in supporting, connecting, and educating patients and caregivers, and providing a platform for PH advocacy, awareness, and research. Finally, more focus should be placed on the patient perspective in PH research and clinical development.

## AUTHOR CONTRIBUTIONS

**Kirsten Braley:** conceptualization, writing - review & editing. **Kathleen Richardson:** conceptualization, writing - review & editing. **Lenise Whitley:** conceptualization, writing - review & editing. **Katie Werner:** conceptualization, writing - review & editing. **Linda Appleby:** conceptualization, writing - review & editing.

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## CONFLICT OF INTEREST STATEMENT

**K. Braley** would like to disclose that the views she has expressed are her own and not necessarily those of her employer, the Robert Wood Johnson Foundation, which was established as a national philanthropy in the early 1970s with Johnson & Johnson stock, and which is independent from the company. **K. Richardson** has nothing to disclose. **L. Whitley** has nothing to disclose. **K. Werner** is an employee of Team PHenomenal Hope, Inc. **L. Appleby** was an employee of Team PHenomenal Hope, Inc. at the time of manuscript development and is now an employee of Lutheran Hour Ministries.

## ETHICS STATEMENT

Ethical approval was not required for this review.

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