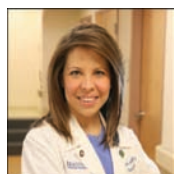


# Quality of Life in Pulmonary Arterial Hypertension: Qualitative Insights From Patients and Caregivers



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Pulmonary arterial hypertension (PAH) is a progressive and presently incurable disease resulting in distressing and debilitating symptoms for patients, including exertional dyspnea, fatigue, chest pain/discomfort, and feeling dizzy or lightheaded.<sup>1,2</sup> These symptoms cause profound functional limitations that often require patients to make significant lifestyle changes and cope with formidable psychological challenges as they face this progressive disease. While the introduction of new treatments over the past decade has expanded options for improving functional exercise capacity,<sup>3</sup> we still have limited insight into the nature of the psychological challenges facing patients and the secondary impact on their caregivers.

We do know that anxiety and depression are common comorbidities in patients with PAH. Data from the multicenter US-based REVEAL Registry showed that clinical depression, defined as patients with the comorbid condition clinical depression and/or patients with the reported use of selective serotonin reuptake inhibitors as a concomitant medication, was present in 25.5% of PAH patients.<sup>1</sup>

This is consistent with other studies: White et al reported a 26% prevalence of moderate to severe depressive symptoms and 19% prevalence of severe anxiety in patients with PAH.<sup>4</sup> Lowe et al noted major depressive disorder in approximately 16% of patients.<sup>5</sup> These publications suggest that depression is substantially more common than the estimated prevalence of major depressive disorder in the general population, estimated at 5%-6%.<sup>6</sup> Beyond depression and anxiety, however, there are clearly other cognitive sequelae affecting patients with PAH, including problems with memory, which has been linked to decreased quality of life (QoL).<sup>4</sup>

While a comprehensive and detailed overview of health-related QoL was recently published in this journal,<sup>7</sup> the remainder of this article will focus on the practical influence that PAH has on the affected patients and their caregivers. Much of this knowledge comes from qualitative research, specifically

several small symptom-experience studies as well as ongoing surveys of PAH patients and their caregivers in collaboration with the pulmonary hypertension (PH) associations in the United States and in Europe. While this work is still in its preliminary stages, there are already some valuable insights that will interest patients newly diagnosed with PAH, their caregivers, and medical providers and associations who strive to optimize the QoL for those affected by PAH.

## QUALITATIVE RESEARCH: PATIENT SYMPTOM EXPERIENCES

Qualitative research has a decidedly different approach than the quantitative studies that have dominated the PAH literature in the area of QoL. Qualitative studies often rely on in-depth, open-ended interviewing techniques and videotaping of subjects to discern the important issues related to the subject matter. The goal is not to prove or disprove a theory at the outset of the work, but rather to develop theories during the process and then use more specific interview questions as the research proceeds. This qualitative approach may convey the “richness of the patient experience”<sup>7</sup> in a way that quantitative surveys cannot generally attain. At least 3 nursing studies have investigated the

patients’ perception of their QoL in the setting of PAH by focusing on qualitative aspects in their research.

The first of these studies by Peloquin et al suggested that prostacyclin therapy may not improve patients’ QoL<sup>8</sup>; however, subsequent research, directly studying patients’ perceptions, refuted these results.<sup>7</sup> In another study, Flattery et al employed semistructured interviews to investigate patient perceptions of living with PAH, and reported 2 overarching themes in their study of 11 patients: patients with PAH suffer from thoughts of uncertainty regarding the future but also tend to overcome these feelings of uncertainty and learn to cope with the illness.<sup>9</sup> Finally, McDonough et al found that “holding back” and “redefining life” were 2 important themes that emerged from patient experiences.<sup>10</sup> Holding back was further described in terms of 3 subthemes: fear, anticipation of worsening symptoms, and treatment effects; while uncertainty, activity restrictions, and “making the best of it” were identified as components of redefining life. The ability to generalize these results is limited by their relatively small sample sizes but provided support for a qualitative approach to investigating QoL in PAH and guidance for possible larger quantitative studies.

## ONGOING PATIENT AND CAREGIVER SURVEYS

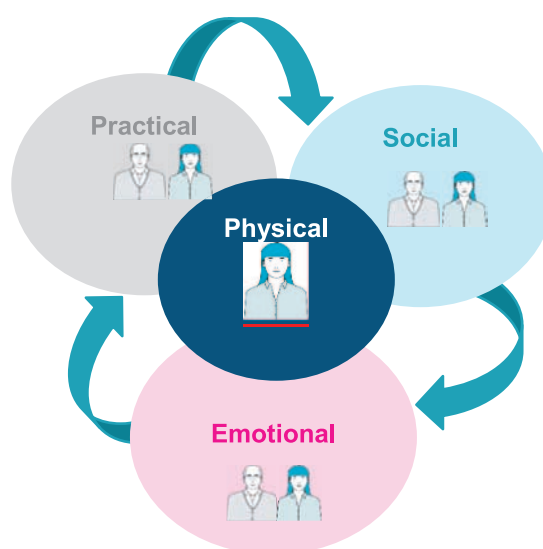
The PH associations in the United States and Europe began collaborating in 2010 to initiate qualitative surveys of patients

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with PAH along with their caregivers.<sup>11</sup> The potential of this work was several-fold: first to determine the influence of PAH on the daily lives of patients and caregivers; second to establish the subject platform for a larger, PAH-specific quantitative QoL survey; and third to provide the PH associations with guidance in their process of developing support programs for patients and caregivers struggling with the effects of PAH. An additional opportunity from this project is the ability to compare responses from Europe with those from the United States to obtain a broader view of the QoL issues across 2 continents.

The United States project, like the one in Europe, was not an academic study sponsored in a university setting but a market research-type, which utilized accepted survey methodologies in a 2-phase project. Phase I involved semi-structured interviews with a small group of patients and caregivers (US sample=19 patients and 12 matched caregivers) and is now completed in both the US and Europe. It was designed to obtain a detailed understanding of the daily challenges facing patients and caregivers affected by PAH, specifically the physical, emotional, social, and practical effect via detailed, first-person characterizations. The effects of depression and attitudes toward seeking information in managing this disease were additional areas of focus. Phase II was subsequently designed as a questionnaire-based survey of a larger sample (sample size of approximately 450 in Europe) to allow for quantification of the Phase I results. Recruitment for this project in the US occurred via outreach efforts by the Pulmonary Hypertension Association (PHA-US) as well as through PH centers in an effort to minimize potential bias of including solely active PH association members.

The interviews in Phase I were recorded and typically lasted 1-2 hours. Most patients were classified as World Health Organization (WHO) functional class II and III, age ranges spanned from 35-75 years (one outlier was 15 years old), and all current US FDA-approved therapeutic classes were represented. The patients were predominantly WHO Group



Patients and caregivers are both **victims** of the **far-reaching** implications and **consequences** of PAH

**Figure 1: Major themes identified in Phase I interviews of patients and caregivers regarding impact of PAH.**

1, although one identified chronic thromboembolic PH (WHO Group 4) as a diagnosis.<sup>11</sup>

While the analysis of the Phase I data in the US and Phase I and II data in Europe is still ongoing and more detailed information is forthcoming, some initial key insights from Phase I have already emerged that are worthy of brief discussion. The results are covered under the themes of **physical**, **social**, **practical**, and **emotional** impact (Figure 1), and the italicized words represent language commonly employed by the interviewees.

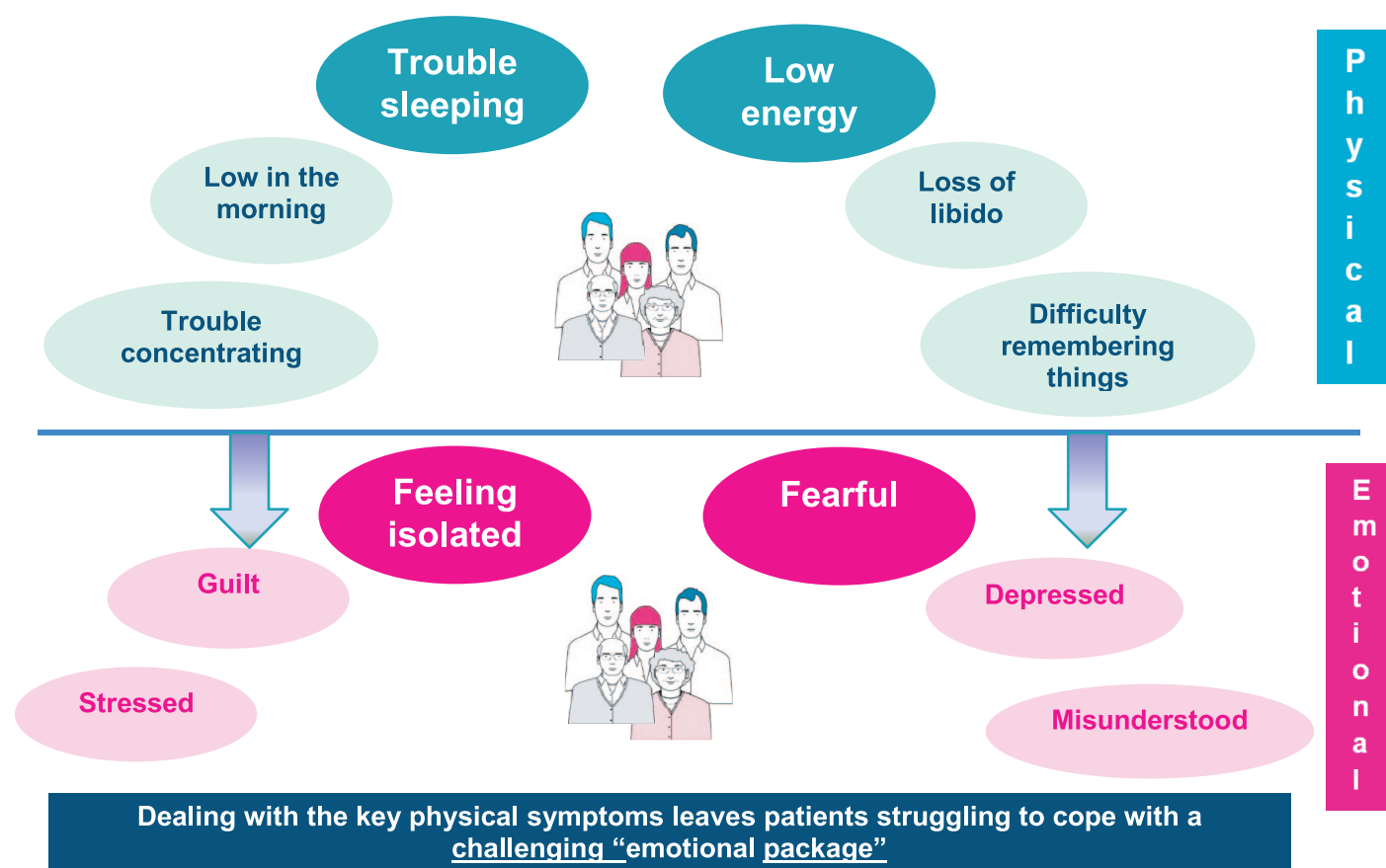
Both patients and caregivers involved in the US Phase I described PAH using vivid imagery (Table 1) as an illness of far reaching consequences that affected many aspects of their lives. They reported physical symptoms that were often *distressing* and *debilitating*, with the result of shifting

practical tasks such as housework to their caregiver. **Physical** limitations also affect patients and their caregiver's ability to *continue to work* and create concerns regarding *finances*. Physical limitations overlap with overall **social** life, which generally suffers due to low motivation and energy, challenges presented by traveling, and feelings of isolation due to the lack of awareness of the condition. Patients described being acutely aware of being *stared at*. There was a strong sense that emerged during the Phase I interviews that the general population has a very limited understanding of PAH, and patients often felt misunderstood. One patient characterized the situation by saying, "Why would anybody want to be my friend? I'm a freak."

The **practical** issues overlay many of these physical and social concerns. Care-

**Table 1: Language Employed by Patients and Caregivers to Describe Their Experience With PAH**

Patients	Caregivers
<i>Breathing through a straw</i>	<i>Having a third job</i>
<i>Being grounded</i>	<i>Always being on call</i>
<i>Living with a time bomb</i>	<i>Not knowing what the future will hold</i>
<i>An ongoing battle</i>	<i>Walking a tightrope without a safety net</i>
<i>Having a ball and chain that never goes away</i>	<i>At times overwhelming</i>



**Figure 2: Physical symptoms suffered regularly by patients, bring with them significant emotional impact.**

givers in particular noted that they were required for *round-the-clock care* and worried about leaving the PAH patient alone for any period of time. They had increased responsibility for *ensuring that patients took their medications* and for *time management* in completing daily tasks, as the patients often lost their motivation and energy. Patients sometimes felt a loss of independence: “They treat me like I’m a baby.”

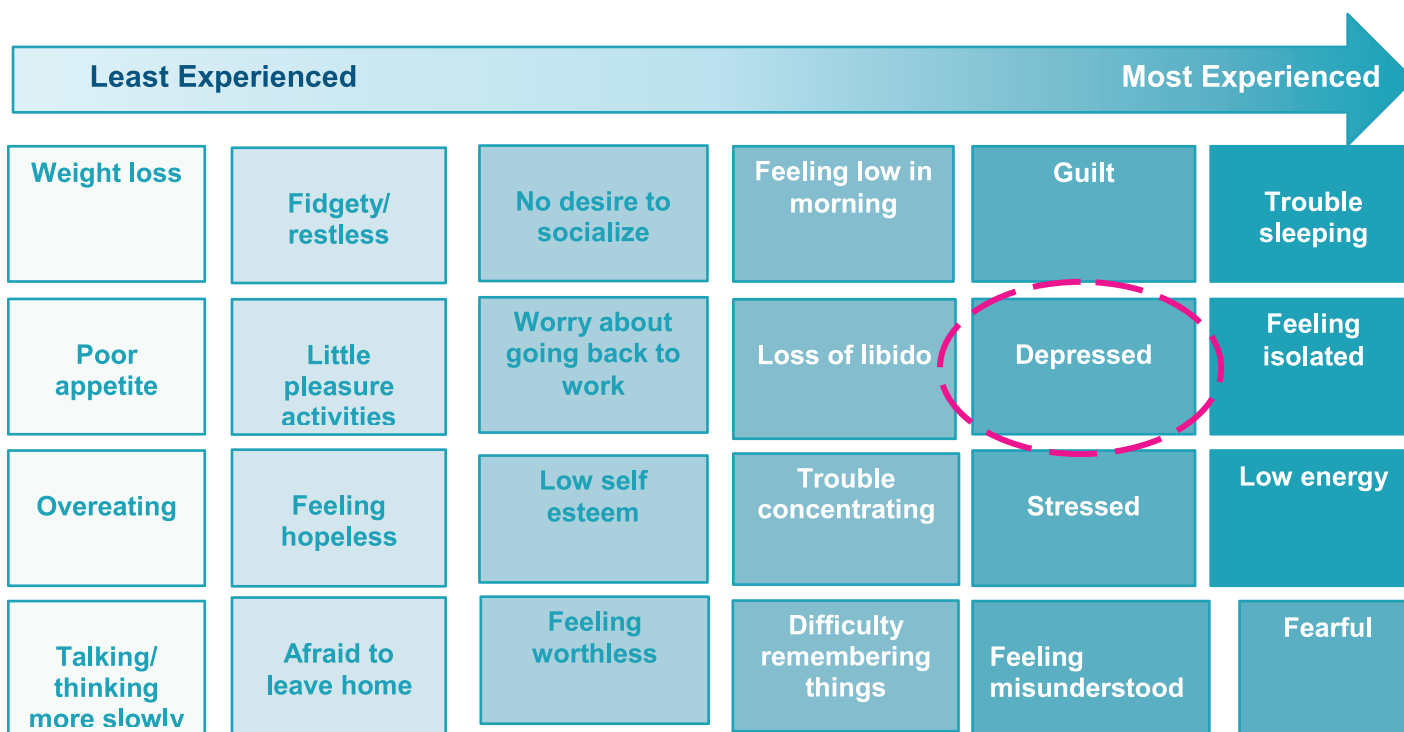
The **emotional** effects of PAH covered a wide spectrum, with some patients describing very negative emotions, such as *sadness*, *anger*, and *shock*, while also experiencing positive emotions including *gratitude at being alive* and benefiting from *increased support* from loved ones. *Uncertainty* regarding the future was a recurrent subtheme and feelings of *guilt* at not being able to continue their traditional roles in society and/or their family. Dealing with uncertain *finances* as well as potential feelings of *resentment* and frustration had a nega-

tive secondary impact on patients’ health status. These issues strengthened the patient-caregiver bond in some instances, whereas in others the burden and pressure of PAH altered the relationship dynamics and had a negative effect on intimacy. Patients and caregivers noted that health care providers (HCPs) did not proactively address their symptoms, and even when they raised the issue with their HCPs many expressed their feelings of dissatisfaction with the HCPs’ responses. Patients also recognized the interaction between their physical limitations and their emotional responses, as summarized in Figure 2.

As noted, depression is a common comorbid condition in patients with PAH; both patients and caregivers recognized the condition itself, as well as clear symptoms of depression. Caregivers often noted depression symptoms in the patients as well as themselves. In a card sorting exercise where the cards contained various classic signs of depression, patients

most often felt fearful, isolated, and less energetic, whereas weight and appetite loss were less commonly reported (Figure 3).

Surveying patient and caregiver attitudes toward information seeking revealed that widely varying information is provided at the time of diagnosis. Some were provided cold, factual information that led to feelings of fear, whereas others were informed in a much more in-depth consultation session, which was greatly appreciated. The majority gained a functional grasp of their heart and lung problems with PAH, and in researching that information formed opinions regarding sources. The summary of the group’s rating suggested that the Internet tended to be the least reliable while their PAH specialist, other PAH patients, and PAH support groups were the most reliable (Figure 4). Interest in support groups was mixed, however, with some patients describing a sense of community (one respondent: “It was like a family reunion”), whereas others found shared negative experiences to



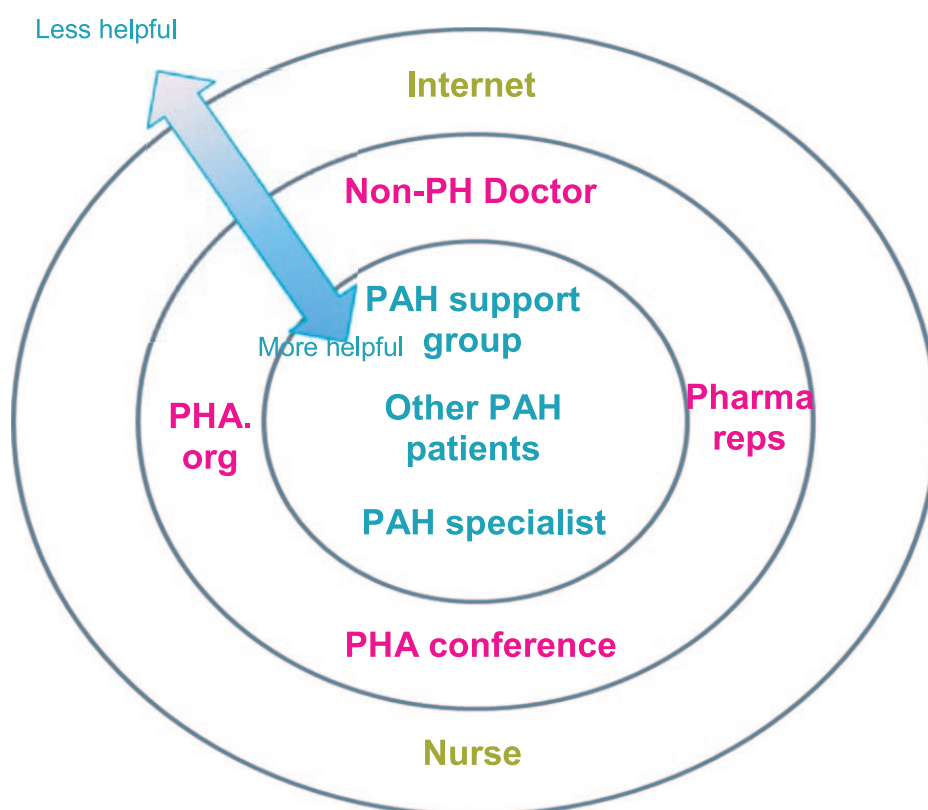
**Figure 3: Results of card sorting exercise demonstrating most common effects of PAH and depression in particular.**

be more depressing than helpful. Almost universally the patients and caregivers expressed their appreciation for their PAH societies' ongoing role in supporting them while they cope with their illness.

These qualitative surveys, which are among the first to solicit the opinions of caregivers along with the patients affected by PAH, have provided some valuable insights into shared life experience as they pertain to QoL. The ongoing Phase II surveys in the US and Europe will further expand this knowledge base. This will hopefully allow HCPs, PAH support groups, and PH societies to help patients and their loved ones better navigate the course of PAH.

## CONCLUSION

A diagnosis of PAH often leaves patients feeling shocked, but this is only the beginning of a long and sometimes arduous journey where patients and their caregivers have to adapt their lifestyles to live with this progressive disease. Understanding the physical, emotional, social, and practical burdens faced by patients with PAH and the secondary impact on their caregivers provides an opportunity for



**Figure 4: Patients' opinions regarding disease information sources. (Abbreviations: PAH=pulmonary arterial hypertension; PHA= Pulmonary Hypertension Association; Non-PH doctor=nonspecialist in PH; Pharma reps=pharmaceutical company representatives).**

those who provide support and care to be more effective. Qualitative research, as well as patient and caregiver surveys related to QoL in PAH, will provide that understanding and will direct ongoing quantitative research and survey efforts with the ultimate goal of improved quality of life for patients and their caregivers.

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