Patients’, relatives’, and practitioners’ views of pulmonary arterial hypertension: A qualitative study

Sophie Alami¹, Vincent Cottin², Luc Mouthon³, Dominique Desjeux⁴, Esther Quessette⁵, Serge Poiriaudeau⁶, Olivier Sitbon⁷

Introduction > To study practitioners’, patients’ with PAH, and relatives’ views regarding pulmonary arterial hypertension (PAH) and identify potential improvements in medical care strategies.

Methods > A qualitative study based on semi-structured interviews involving 16 patients, 4 relatives, and 9 practitioners.

Results > Patients with PAH, their relatives, and physicians who treat persons with PAH had divergent perspectives on PAH. The discrepancies identified concerned their perceptions of the illness and its impact on patients’ with PAH daily lives. Patients with PAH had a broader view, including social, identity, financial, and functional dimensions of PAH’s impact on their lives, whereas practitioners were more focused on functional aspects. The study also pointed out divergent approaches among physicians to assessing patients’ New York Heart Association functional class. The expectations of patients with PAH, relatives, and physicians also differed. Patients with PAH expected improvement in PAH diagnosis and better coordination between primary care physicians and PAH medical centers. They also valued reducing side effects, less restrictive medications, and greater consideration of their views in the medical decision making process. Physicians’ expectations focused more on identifying and validating therapeutic strategies.

Conclusion > Our results suggest several potential improvements in patient management, especially in order to better tailor treatment to patients’ needs and to achieve a more uniform approach.
Pulmonary arterial hypertension (PAH) is a rare fatal disease characterized by vasoconstriction, in situ thrombosis, and vascular remodeling of small pulmonary arteries leading to increased pulmonary arterial resistance, right heart failure, and eventually death [1]. The French registry of patients with PAH [2] provides an estimate of PAH prevalence in France of 15.0 cases/1 million adult inhabitants. This registry identified a female-to-male ratio of 1.86 and 1.32 for the incident cases, and a distribution of PAH patients according to New York Heart Association (NYHA) functional classification of 1% in class I, 24%

Résumé

Regards croisés sur l'hypertension artérielle pulmonaire : une étude qualitative réalisée auprès de médecins, de patients et de leur famille

Introduction > Le point de vue et les attentes des personnes atteintes d'hypertension artérielle pulmonaire (HTAP), de leurs proches et des médecins qui les prennent en charge a jusqu'ici été très peu étudié.

Objectif > Apprêcher le point de vue des médecins, des personnes atteintes d'HTAP et des membres de leur entourage sur la maladie et sa prise en charge et identifier des pistes d'amélioration des stratégies de prise en charge.

Méthode > Une étude qualitative a été conduite auprès de 16 personnes atteintes d'HTAP, de 4 membres de leur famille et de 9 médecins.

Résultats > Interrogées sur leurs représentations de la maladie et son incidence sur leur vie quotidienne, les personnes souffrant d'HTAP ont évoqué des dimensions qui apparaissent négligées par les médecins. Alors que le discours des praticiens était centré sur la seule dimension fonctionnelle, les personnes atteintes d'HTAP ont également souligné l'importance de l'incidence sociale, identitaire et financière de la maladie. De même, tandis que les attentes des médecins portaient sur l'identification et la validation de stratégies thérapeutiques, celles des patients concernaient : la réduction des délais diagnostiques ; une meilleure coordination entre les médecins de soins primaires et les centres médicaux spécialisés ; l'amélioration de l'efficacité thérapeutique ; la diminution des effets secondaires ; une offre de traitements moins contraignants ainsi qu'une amélioration des modalités de leur participation aux décisions médicales les concernant. Enfin, l'examen des pratiques d'évaluation de l'incidence fonctionnelle de l'HTAP a permis de mettre en lumière l'hétérogénéité des pratiques médicales d'évaluation de la classe fonctionnelle des patients selon les critères de la New York Heart Association.

Conclusion > L'étude met en lumière l'écart de perspective qui peut exister entre patients et praticiens. Elle incite à une meilleure prise en compte des objectifs, des priorités et des préférences des patients dans l'adaptation des traitements. Elle souligne l'hétérogénéité des processus d'évaluation de l'incidence fonctionnelle de l'HTAP au sein des praticiens. Elle constitue une source d'information susceptible d'enrichir l'éducation thérapeutique à destination des patients et de leurs proches et de permettre aux praticiens de mieux interpréter la dyspnée chez les personnes atteintes d'HTAP. Enfin, les résultats de cette étude constituent une base de données qualitative permettant d'identifier les critères d'évaluation rapportés par les patients nécessaires au développement de nouveaux instruments (patient reported outcome measures) permettant d'enrichir l'évaluation de l'impact de l'HTAP sur la vie quotidienne des personnes malades.
in class II, 63% in class III and 12% in class IV [2]. Despite significant progress over the past decade, PAH remains incurable [3, 4]. The 1-, 2-, and 3-year survival rates are respectively 87%, 76%, and 67% [5]. In addition, PAH results in impaired quality of life [6–10] especially due to dyspnea and severe functional limitations. The relevance of patients’ perspectives to the medical decision-making process and product development has been pointed out [11–15], and guidelines have been edited that broaden the traditional perspective of medicine by considering indicators of what “really matters” for patients as part of therapeutic assessment [16]. The recent focus on patient-reported outcomes (PROs) emphasizes the growing recognition that patients’ perspectives are relevant to therapeutic issues [10–17]. This patient-centric shift makes improvement in patients’ quality of life a part of healthcare professionals’ concern, along with biological and clinical goals [13]. Listening to the patient’s voice for more than the mere detection of symptoms may be beneficial to healthcare.

Little is known about patients’ views of PAH and their expectations. A qualitative approach can help explain patients’ context and needs, determining what benefits they seek beyond healing, and identifying how they define their “quality of life” and their therapeutic goals [18]. However, research focusing on patients’ perspectives has been minimal, and no study was found comparing patients’, relatives’, and physicians’ perspectives regarding PAH. Two previous qualitative studies [7, 19] and 2 literature reviews [6, 10] have addressed the issue of health-related quality of life (HRQoL) in PAH. Three qualitative studies have addressed patients’ perspectives in PAH [20–22], all of them focusing on exploring only the patients’ experience of PAH. Combining patients’, relatives’, and practitioners’ views provides a comprehensive picture of PAH, enriching medical knowledge with patients’ concerns, goals, and expectations, which are all at the heart of the therapeutic contract, a prerequisite for improving shared decision making. The objective of the study was thus to explore practitioners’, adults’ with a diagnosis of PAH, and their relatives’ views regarding PAH and to identify potential improvements in medical care strategies. In this perspective, this study has begun the work of identifying and standardizing PROs in PAH [10], a preliminary step in developing new PROs. There were three groups of study participants: “patients with PAH” (or “patients”), “relatives”, and “physicians”. Our research questions were:

• What are patients’, relatives’, and physicians’ views of the disease, of the limitations, and of quality of life in PAH?
• Are there any differences among the groups’ views and priorities?

Methods

Ethics statement

The study protocol was approved by the local ethics committee, the “Comité de protection des personnes Île de France III of Tarnier – Cochin Hospital”, Paris. All participants gave written informed consent to participate in the study [more details provided in supplementary file S1]. Investigations were conducted according to the Declaration of Helsinki principles.

Design

To address the goals of this study, given the limited existing research on the topic, a qualitative research strategy was developed to deal with its exploratory, comprehensive, and descriptive concerns, and the study was conducted in accordance with qualitative guidelines [23–29]. An inductive enquiry consistent with the grounded theory approach [25] was adopted: the study sought to provide a rich description of PAH experience from the perspectives of 3 major groups of PAH stakeholders, to distinguish their needs and concerns, and to identify differences in order to improve patient management.

Participants and sampling

The sample selection was based on a non-probability judgment sampling, seeking both relevance to the subject of the study and diversity among the individuals selected. We opted for purposeful sampling in order to maximize heterogeneity to provide rich and diverse data relevant to the research question. Patients with PAH were recruited at the French National Center for Severe Pulmonary Hypertension, Department of Pneumology and Intensive Care, hôpital de Bicêtre, Le Kremlin-Bicêtre, France. The sample size was determined on the basis of the principle of maximum variety sampling. To explore diversity, gender, age (over 18 years old), functional impairment and living environment served as starting points for sampling. However, to clearly circumscribe the results to PAH, individuals with PAH associated with other diseases were excluded. Saturation [25] was only a guiding principle. This concept can provide a sample size only contingently (i.e., contingent on data collection and analysis, as per Glaser and Strauss). Therefore, we relied on the professional experience and competency of the research team to determine sample size. To ensure the diversity of the physician sample, we recruited doctors with expertise in PAH from different specialties (pulmonologists, cardiologists, and internists) and from different medical departments throughout the country. To be eligible, relatives had to be free of PAH and to be part of the patients’ daily life. Diversity was obtained by selecting a variety of relations and male and female relatives.

8 Saturation occurs in data collection when informational redundancy appears. No new findings emerge; new data no longer bring additional relevant information to the research questions.
Recruitment
To maximize the diversity of participants, recruitment was done by 3 different informants working at the National Reference Center for Severe Pulmonary Hypertension. Patients who participated in the study were approached by 2 physicians (one of whom was involved in the study) and a nurse in charge of PAH patient therapeutic education. Those doctors also recruited 2 relatives, the other 2 being recruited through a snowball strategy with the help of patients with PAH who were interviewed.

Data collection procedure
Data were collected by 2 sociologists. The physician interviews were conducted during winter 2010 and interviews with patients with PAH and relatives from April to July 2011 [S2]. Interview protocols were developed to offer an open framework for focused but flexible data collection [S3]. The interviews were fully transcribed [S4]. All interviews were conducted with a commitment to respect the anonymity of respondents.

Analysis
The data set containing all the written transcripts was analyzed by the sociologists who conducted the interviews. We read a series of interviews and inductively developed an initial list of themes. This first list provided a common content analysis grid for the entire corpus. Throughout the thematic analysis, this list was expanded, made more detailed, and modified as we found new insights about the data. We organized sessions of peer debriefing with PAH specialists and a third sociologist to ensure high levels of inter-researcher consistency in analysis, and we discussed our interpretations of the data to ensure accuracy and enhance the validity of the analysis.

Results
Sample characteristics
A sample of 16 patients with PAH (6 men), 4 relatives (2 men), and 9 physicians (8 men) were interviewed. Patients’ medical characteristics and personal and professional status are displayed in Table I. Patients were aged between 24 and 75. Twelve were idiopathic PAH and 4 heritable. Six were NYHA functional class II, 8 were class III, and 2 were class IV. Out of 16 patients, 7 lived in the Paris area and 9 in other areas, including 3 in rural areas. The relatives interviewed were 1 wife, 2 husbands, and 1 sister who lived together with the patient (2 men; 2 women), all of them living in the Paris area (table II).

<table>
<thead>
<tr>
<th>Patient Number</th>
<th>Sex</th>
<th>Age</th>
<th>NYHA class</th>
<th>Etiology</th>
<th>Occupation</th>
<th>Current work status (at interview date)</th>
<th>Family status</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>Female</td>
<td>64</td>
<td>III</td>
<td>Idiopathic</td>
<td>Teacher</td>
<td>Retired</td>
<td>Single</td>
</tr>
<tr>
<td>P2</td>
<td>Female</td>
<td>30</td>
<td>II</td>
<td>Idiopathic</td>
<td>Accountant</td>
<td>Working</td>
<td>Single</td>
</tr>
<tr>
<td>P3</td>
<td>Female</td>
<td>57</td>
<td>II</td>
<td>Heritable</td>
<td>Housekeeper</td>
<td>Retired</td>
<td>Married</td>
</tr>
<tr>
<td>P4</td>
<td>Female</td>
<td>66</td>
<td>II</td>
<td>Idiopathic</td>
<td>Secretary</td>
<td>Retired</td>
<td>Widow</td>
</tr>
<tr>
<td>P5</td>
<td>Female</td>
<td>42</td>
<td>IV</td>
<td>Idiopathic</td>
<td>Secretary</td>
<td>Non-working</td>
<td>Single</td>
</tr>
<tr>
<td>P7</td>
<td>Female</td>
<td>66</td>
<td>III</td>
<td>Idiopathic</td>
<td>Housewife</td>
<td>Non-working</td>
<td>Widow</td>
</tr>
<tr>
<td>P10</td>
<td>Female</td>
<td>30</td>
<td>III</td>
<td>Heritable</td>
<td>Caregiver</td>
<td>Non-working</td>
<td>Single mother</td>
</tr>
<tr>
<td>P13</td>
<td>Female</td>
<td>51</td>
<td>III</td>
<td>Idiopathic</td>
<td>Housewife</td>
<td>Non-working</td>
<td>Separated</td>
</tr>
<tr>
<td>P14</td>
<td>Female</td>
<td>30</td>
<td>III</td>
<td>Idiopathic</td>
<td>Ambulance attendant</td>
<td>Non-working</td>
<td>Single</td>
</tr>
<tr>
<td>P15</td>
<td>Female</td>
<td>42</td>
<td>III</td>
<td>Idiopathic</td>
<td>Sound technician</td>
<td>Non-working</td>
<td>Single</td>
</tr>
<tr>
<td>P6</td>
<td>Male</td>
<td>51</td>
<td>II</td>
<td>Idiopathic</td>
<td>Computer engineer</td>
<td>Working</td>
<td>Married</td>
</tr>
<tr>
<td>P8</td>
<td>Male</td>
<td>63</td>
<td>IV</td>
<td>Idiopathic</td>
<td>Research manager</td>
<td>Retired</td>
<td>Single</td>
</tr>
<tr>
<td>P9</td>
<td>Male</td>
<td>24</td>
<td>III</td>
<td>Heritable</td>
<td>Policeman</td>
<td>Working</td>
<td>Married</td>
</tr>
<tr>
<td>P11</td>
<td>Male</td>
<td>75</td>
<td>III</td>
<td>Idiopathic</td>
<td>Store manager</td>
<td>Retired</td>
<td>Married</td>
</tr>
<tr>
<td>P12</td>
<td>Male</td>
<td>42</td>
<td>II</td>
<td>Heritable</td>
<td>Agriculture worker</td>
<td>Non-working</td>
<td>Married</td>
</tr>
<tr>
<td>P16</td>
<td>Male</td>
<td>52</td>
<td>II</td>
<td>Idiopathic</td>
<td>Legal expert</td>
<td>Working</td>
<td>Cohabiting</td>
</tr>
</tbody>
</table>
The 9 doctors interviewed worked in 8 different locations: Caen, Clamart, Lille, Lyon, Marseille, Nancy, Paris, and Strasbourg.

Patients' views of PAH

The representation of the illness: a complex disease difficult to decipher

For patients with PAH, the illness comes as a new disease, and they must devise a representation of it. To begin with, the full name of the disease is problematic for them:

"(What do you call it?) PAH. I had a hard time to get my head around it! But it is in there now. I say "PAH" and that's it!" (patient 3)

PAH is a confusing label for them. The presence of the word "hypertension" may obscure the specificity of the disease, evoking systemic hypertension, a more common disease associated with less worrisome representations. However, the representation of PAH is progressively shaped by combining information provided during the patients’ medical journey. The disease itself is difficult to understand, especially regarding the physiological mechanisms involved. Patients had problems defining the territory affected by the disease. They did not identify the pulmonary artery as the center of the disease, and they failed to clearly describe the mechanisms of the disease; rather, they mentioned various phenomena (i.e., narrowing, thickening, hardening, contraction, or cell proliferation) that disrupt blood flow and gas exchange, and eventually deteriorate their heart:

"PAH, well, high pulmonary artery pressure. It affects the heart and lungs, the liver, the kidneys. All that's mixed around it! But it is in there now. I say "PAH" and that's it!" (patient 3)

"It's the blood that goes... it's my artery... the blood that doesn't... hold on... it's the blood that goes from the heart to the artery... I don't know how to explain it. I'm afraid I... (No, don't worry. I'm not an expert.) They've explained it to me several times, but..." (patient 10)

"It's complicated. It definitely is complicated. I'm amazed, because there are patients who know it by heart. They know all the scientific terms. They're learning medicine on their own at home! (laughs)" (patient 16)

In describing PAH, patients also emphasized the rarity of the disease, as well as its incurability and its impact on life expectancy:

"(If I asked you to explain the disease, PAH... what could you tell me?) Not much. It's an awful disease. You die. That's all. And it always gets worse. It never gets better." (patient 3)

The rarity of the disease means that the general public knows nothing about it, and health professionals are not always familiar with it:

"Society doesn't know about it. It's not visible. It's just one more disease. It's there for those who know about it, that's it! No more details. If it were something that more people knew about, like the diseases you hear about on TV, but there aren't many of us." (patient 2)

"They tell us afterwards that for a general practitioner, there's less than one chance in a hundred that he'll come across PAH in his career." (patient 6)

Profane semiology of PAH

Patients characterized the disease by describing the symptoms that they perceived and the impact of the disease on their daily lives. They cited 19 symptoms (Table III), not all of which are clearly associated with their illness. For some symptoms, the link with the physiological mechanisms of the disease is not clear to them (weight loss, cramps, feeling of nausea, dry cough); others resemble certain side effects of medications (leg pains). Some are not mentioned by doctors during consultations (fatigue).

Patients identified 2 categories of symptoms. The first category includes symptoms that are short-lived and recurrent, occurring after an activity. Patients mentioned exertional dyspnea, thoracic pain, dizziness, syncope, liver pains, palpitations, "spitting blood," Raynaud’s phenomenon, cyanosis, cramps, leg pains, "dry cough," and dyspnea at rest. These symptoms are viewed as bodily reactions triggered by particular activities or contexts. The second category consists of symptoms that become long-lasting and are felt throughout the day, for which patients do not identify specific triggering events or contexts. Patients cited

<table>
<thead>
<tr>
<th>Relative number</th>
<th>Sex</th>
<th>Age, years</th>
<th>Occupation</th>
<th>Relation to interviewee</th>
</tr>
</thead>
<tbody>
<tr>
<td>R1</td>
<td>Female</td>
<td>54</td>
<td>Engineer</td>
<td>Wife</td>
</tr>
<tr>
<td>R2</td>
<td>Female</td>
<td>39</td>
<td>Pre-school teacher</td>
<td>Sister, sharing home</td>
</tr>
<tr>
<td>R3</td>
<td>Male</td>
<td>59</td>
<td>Surveyor</td>
<td>Husband</td>
</tr>
<tr>
<td>R4</td>
<td>Male</td>
<td>35</td>
<td>Technician</td>
<td>Husband</td>
</tr>
</tbody>
</table>

The 9 doctors interviewed worked in 8 different locations: Caen, Clamart, Lille, Lyon, Marseille, Nancy, Paris, and Strasbourg.

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insomnia, heightened sensitivity, lack of appetite, and weight loss. Fatigue is classified in both categories; it is viewed as a permanent symptom that may become more severe in specific situations.

**Dyspnea: a major symptom with differing forms**

No matter which other symptoms they mentioned, patients identified breathlessness as the primary symptom of PAH. Although they called it “the main symptom” because of its frequency, it was nevertheless difficult for them to describe. Patients used different registers to describe it, showing multifaceted feelings, ranging from simple discomfort to pain. Three aspects intersect: sensory perception, functional impact, and emotions, related to the fear that they feel. With dyspnea, breathing becomes a conscious act:

“[Shortness of breath is] when you breathe and you become aware of your breathing, that it isn’t normal. When you breathe, you do it unconsciously. Running out of breath is like saying “Aha!” As I said, when I do things calmly, I don’t realize I’m doing them. I’m not aware.” (patient 1)

Then they described it as a “little problem” whose intensity and form may vary. It may be associated with difficulty in breathing, a feeling of choking or suffocating, oppressiveness or crushing chest pain, or severe pain. Fear and suffocation may also combine to create a feeling of “suffering”:

“What do I say so that people understand? When you do sports and you’re short of breath and you’re normal, you know... shortness of breath is effort. It’s an effort, it’s an inconvenience. When you don’t breathe well because of a disease such as PAH, it’s not an inconvenience, it’s a pain. It’s not on the same level of pain – well, you’re suffering... It’s painful. There are signs that it’s painful in your armpits. Around your shoulders. It feels... widespread. It’s not just a failure, and you can’t provide oxygen. It’s something else. There are other parts of the body that do not follow. It’s something else. And really, it isn’t effort; it’s pain. It’s not the same thing.” (patient 6)

Patients did not always clearly identify the specific contexts in which shortness of breath occurs. Some mentioned a feeling of...
breathlessness not caused by any physical activity, during sleep or rest, sitting or lying down. Dyspnea was described by patients as resulting from physical movements, whether requiring an effort or not; from other symptoms, such as palpitations, cough, edema or fatigue; from environmental conditions such as extreme temperatures, humidity, or pollution; or from an emotional shock.

When designating specific activities that trigger dyspnea, each interviewee mentioned a variety of contexts, depending on the severity of the disease (tables IV and V).

The impact of PAH: life disrupted

The disease was a disruption in the lives of patients with PAH. Patients gradually discovered the effects of the disease on their daily lives and became aware of the limitations that it imposed. It redefined their lives and their vision of the future. There were multiple implications: functional, financial, identity, and social. The physiological and physical consequences of PAH impact all areas of the patients’ lives. They affect, in functional terms, household chores, professional activities, leisure activities (sports, travel, shopping, cultural activities), body care and dressing, parenting, sexuality, and mobility. Patients were forced to reduce or eliminate activities:

“For 6 months, I stayed home, hooked up to oxygen. So no work, no housework; I wasn’t allowed to do anything. For the time it took to get back to normal. I was connected 24 hours a day, 7 days a week. I was really bored for those 6 months, just lying there and watching TV. I was at home [in the apartment I share with my sister]... Lots of things have changed, obviously. I do a lot less.” (patient 2)

The idea of effort is reflected throughout the interviewees’ statements when they describe their symptoms and the impact of the disease; often when describing an activity that provoked a symptom. Different terms were used: “for any effort,” “everything becomes an effort,” “a little effort,” “a big effort,” “it is an effort for me.” Effort has different meanings for them, but all patients with PAH reported that they had to restructure their lives. The results showed that PAH alters the conventional perception of effort (table VI). Actions that “normally” do not require much effort (such as “putting on pants/shoes, carrying a...
working folder, or showering") become onerous, and patients may find themselves torn between the social norms that define effort and their own personal perceptions. The symptom becomes the indicator of what is an effort or not. The definition of effort changes over time: it varies according to the fluctuations of the disease and its severity, and more and more activities are involved.

In material terms, the disease can lead to a reduction in income, some patients undergoing major financial difficulties. Finally, in terms of identity, patients described a loss of self-esteem related to the discrepancy between the image they would like to project and the reality of their daily life:

### Table V

**Activities outside the home viewed as causing shortness of breath**

<table>
<thead>
<tr>
<th>Type</th>
<th>Subtype</th>
<th>Activity</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Work</em></td>
<td>Job</td>
<td>Oral presentation</td>
</tr>
<tr>
<td></td>
<td>Work-related efforts</td>
<td>Carrying files, objects</td>
</tr>
<tr>
<td><em>Consumption</em></td>
<td>Shopping</td>
<td>Pull/push the shopping cart, Carrying shopping bags, Loading/unloading trunk</td>
</tr>
<tr>
<td><em>Leisure</em></td>
<td>Sports</td>
<td>Bicycling, running, skiing, tennis, squash, yoga, swimming, hiking, fishing, team sports</td>
</tr>
<tr>
<td></td>
<td>Gardening/do it yourself</td>
<td>Mowing the lawn, weeding, trimming, planting flowers/vegetables, Sawing, moving wood</td>
</tr>
<tr>
<td></td>
<td>Culture/entertainment</td>
<td>Going to movies/concert/museum/theater, Sight-seeing, Carrying luggage</td>
</tr>
<tr>
<td></td>
<td>Friends and family</td>
<td>Going out: discotheque, restaurant, parties, evening celebrations, Visiting friends/family</td>
</tr>
<tr>
<td><em>Movement</em></td>
<td>Walking</td>
<td>Walking in itself, Going to: market, work, hairdresser's, from house to car, Strolling</td>
</tr>
<tr>
<td></td>
<td>Running</td>
<td>Catching a bus/subway</td>
</tr>
<tr>
<td></td>
<td>Climbing</td>
<td>Going up: stairs, curb, slopes, Getting on bus</td>
</tr>
<tr>
<td></td>
<td>Public transportation</td>
<td>Walking to bus/subway/car/train, Standing in public transportation</td>
</tr>
</tbody>
</table>

### Table VI

**Redefining effort**

<table>
<thead>
<tr>
<th>Context of symptom occurrence</th>
<th>Perception of effort</th>
<th>Activities causing dyspnea</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>At rest</strong></td>
<td>No effort identified</td>
<td>Sleeping – lying in bed – watching TV – talking – without any reason</td>
</tr>
<tr>
<td><strong>Regular, short activities</strong></td>
<td>Not viewed as effort before PAH</td>
<td>Getting dressed – bathing – carrying</td>
</tr>
<tr>
<td><strong>Regular, longer activities (&gt;10 min)</strong></td>
<td>Increase in effort perceived</td>
<td>Housework – gardening – shopping</td>
</tr>
<tr>
<td><strong>Exertion</strong></td>
<td>Increase in effort perceived</td>
<td>Sports</td>
</tr>
</tbody>
</table>
"Since I have PAH, yes, I’ve changed a lot. I don’t put on makeup anymore; I don’t keep up my looks. Yes, a lot of things have changed.” (patient 10)

"Afterwards I thought, “I wanted to be a fit retiree.” Well, I guess that’s not working out, either. The way that I saw myself maturing physically has changed.” (patient 6)

The disease affected all daily decisions, from trivial matters such as fetching one’s mail to much more important ones — living alone, moving, having a child, and so on. Patients were continually adapting their actions (table VII).

The journey to diagnosis
The period preceding diagnosis included a gradual construction of symptoms by the patients, in 4 stages. Interviewees reported becoming progressively aware of physical disorders, which they did not immediately decipher as “symptoms” requiring medical care.

At first, patients tended to dismiss their symptoms by blaming shortness of breath, fatigue, and coughing on unusual weather conditions (excessive heat), difficult topography, intensity of the effort needed, or being out of shape. Secondly, patients justified symptoms due to a prolonged physical effort as a sign of a previously diagnosed disease (e.g., asthma) or hereditary condition. Patients did not consider their condition “abnormal,” and they developed systematic coping strategies. Their relatives continued to minimize the importance of the symptoms. In the third stage, patients were confused by symptoms that they could no longer justify. Their symptoms intensified, and new ones appeared, such as edema, dizziness, palpitations, stiffness, choking, and pain. Discomfort turned into disability, and adaptation strategies could no longer provide relief for their symptoms. Finally, new symptoms appeared even at rest and significantly disrupted their everyday life. No explanation sufficed; they required medical assistance and relatives encouraged them to seek it.

The first recourse to medical assistance did not systematically produce a diagnosis of PAH. Patients reported hearing multiple other interpretations of their symptoms. Misinterpretation of symptoms delayed the diagnosis:

“At first it was, “you have stomach flu” and “you’re expecting a baby...” Then, finally, I saw a doctor who was good, who had me do some tests. That’s when they found that I had...” (patient 5)

"[With the sports doctor,] we did blood tests. They saw that I had 18 milligrams per liter of red blood cells. They said there was a problem with the EPO: “You have too many red blood cells. There must be a problem with your kidneys.” (patient 6)

”[My doctor couldn’t figure out what I had]. I saw that it was continuing. In June, in the office, I really felt bad. I felt that everything was wrong. I called my doctor and told him, “Listen, we have to do something. I can’t handle it. He told me, “Well, I’ll make an appointment for you to have a coronary check to see what it is...” [After the coronary exam,] the doctor told me: “Listen, we really don’t understand what you have. Your heart is hardening.” So with my husband... I immediately asked “Why is my heart hardening? How’s that possible? I don’t understand!” He told me: “Neither do we...” I was diagnosed in February or March 2004 – it took quite a while [to obtain a diagnosis]! A whole year! [It took so long because] I don’t think they know much about the disease.” (patient 4)

Therapeutic management and evaluation of PAH
The evaluation of the patient-health professional relationship gave rise to contrasting assessments by patients with PAH in this study, who distinguished between physicians who specialized in pulmonary hypertension and those who did not. Their
relationship with PAH specialists was evaluated in terms of the dialogue they established with their practitioners and the importance they were given in the therapeutic decision-making process. Trust was not granted automatically, and could be withdrawn at any point in the patient's journey. It depended on the amount of time the doctor accorded to discussion with the patient and the degree of commitment that a patient felt. They appreciated doctors who considered their psychological state and way of life, and who regularly explained the progression of the disease and changes in treatment. Patients in this study expressed dissatisfaction at the lack of accessibility and availability of doctors. As for non-specialist physicians in PAH, patients primarily criticized their lack of knowledge about the disease.

"The problem is the lack of qualified doctors. The big problem is that the specialty center is here (and I live in another region). It's very far. . . It's a bother because we always have to call here. We're stuck. The doctors I can see at home don't have enough information." (patient 12)

"(Did they tell you what these tests are?) No. But anyway here, they don't say anything. You do the tests, and you don't find out anything. They put everything in the pouch, and it's 'fend for yourself.' They say: 'You have to take such and such a drug, this and that, and that's it.' And a blood test every month!" (patient 3)

When questioned about their medical care, patients also evaluated their treatments. Six criteria were identified; the first criterion is based on the healing effect. As there is no effective healing treatment for PAH, patients used additional criteria. Efficiency is the second criterion; it is associated with the elimination or alleviation of symptoms, such as the reduction of respiratory recovery time and the recuperation of the capacity to perform certain activities. The third criterion mentioned is side effects. The fourth criterion refers to convenience and ease of use (route of administration, number of required doses, autonomy in administration, schedule, hygiene, and painfulness). The fifth criterion involves safety and the risk of infection. The last criterion points out psycho-social consequences of treatments; subcutaneous, intravenous, and inhaled treatments were cited as creating social stigma and isolation (12 patients out of 16 had experienced epoprostenol, 6 oxygen therapy, and 1 subcutaneous treprostinil). Patients acknowledged periods of neglect in adhering to their treatment, of varying length. Non-compliance was short-lived with regard to PAH-specific medications, lasting at most one day, out of fear of fatal consequences. Symptomatic drug treatments could be neglected for longer periods of time, particularly in the case of diuretics. Patients may feel that they can dispense with them, that the disadvantages outweigh the perceived benefits, or that the prescribed dose is negatively impacting their health:

"I try to take it easy and do what it takes but also juggle to keep my other organs working properly. I'm not the sort to say, "I'm tired, I'm stopping." Swallowing 15 meds a day. . . I take the ones I have to, I've stopped some that have had bad effects on me." (patient 15)

Alternative therapies were also part of the panorama of treatments mentioned by patients. They reported using homeopathy, herbal medicine, massage, hot baths, and hypnotism, some of which can be alternatives to diuretics and analgesics. Apprehensive about imposing too many drugs on their bodies, they value these alternative remedies, considering them effective and less harmful than the drugs prescribed, while presenting few or no side effects.

When asked about their treatments, patients also focused on 2 tests: the 6-minute walking test and right heart catheterization. Patients perform the walking test with different ideas of how the test is designed to work, and because of this, they question its reliability:

"But I see some people who take all the time they want. So I don't think the walking test is all that reliable. Some people could make a bit more of an effort. You can't really rely on the effort test, because there are people who don't try very hard. Once I saw a man who could have done a better job walking. He didn't make any effort on the walking test, so it didn't work. . . The therapists have had to tell me to slow down, but they know that I really want to see how much I can do, so I know. Some people I see don't make any effort at all, so you can't tell." (patient 12)

Patients clearly identified right heart catheterization as the standard medical test to assess their health status and determine their treatment. However, patients were apprehensive about this test because it may indicate the aggravation of the disease and it may be painful.

Finally, some patients recalled being asked a set of questions during medical examinations. In fact, the questions could vary from visit to visit and from doctor to doctor. The questions were most often open, ranging from "How are you?" to more specific questions such as the recurring "How many flights can you climb without running out of breath?" However, not all patients were aware of the existence of the NYHA functional classification [55], the standard medical assessment tool, nor were they able to identify their NYHA functional class.

Patients' expectations regarding medical care

Patients expected a shorter period between symptom onset and PAH diagnosis and imagined better coordination between primary care physicians and PAH expert medical centers. They suggested strategies for doctors to mitigate the shock when announcing a PAH diagnosis (e.g., announcing in stages, having
a relative present (with a patient's consent) at the first announcement, formally encouraging contacts between patients, offering referral to a psychologist or social worker more than just once). As for the prognosis, they expressed a tension between the need for information in order to "take necessary measures" to "prepare for the worst" and the need to keep up hope.

Whereas all patients wished that a curative treatment would be discovered, they did value the reduction of side effects and less restrictive medications. They also asked for greater consideration of their views in deciding treatment goals and therapy.

**Physicians' views of PAH**

A complex disease with many unknowns

When doctors described PAH, they emphasized that it is rare and difficult to detect. They cited the lack of symptoms in the early stages of the disease and the absence of specificity when they do start to appear. The impact of the disease on the right heart is thus described as the only sure sign of the disease. Not visible to the patient, this sign is identifiable only after 2 specific medical examinations, echocardiography and right heart catheterization:

"There's the rareness on one hand and then there's the exclusive nature of the screening exams on the other, that is to say, the ultrasound of the heart and the evidence adduced by right heart catheterization. [Today] there are more and more diseases for which you need a specific, invasive exam to provide evidence and to make a relatively sure diagnosis." (physician)

Their prognosis was generally pessimistic, the disease remaining incurable and fatal:

"Today, there is an increasingly rich therapeutic arsenal, which was not the case a decade ago. [The patients] have more molecules available. Unfortunately, none offer curative powers." (physician)

However, their representation of the progression of PAH was contradictory; the disease was described as definitely fatal in the short term, but at the same time, medical progress is expected to transform it into a chronic disease:

"What is important is that patients with PAH still die, but in the idiopathic forms, they die less than before, and they live longer. Forms associated with scleroderma are the ones that have the shortest time: 3 years on average. Finally, we are now moving towards a chronic disease. Today, the standard for the disease – which we never imagined would become a chronic disease – is AIDS. . . . The Class II and Class III forms can live 10 or 15 years. . . . Although there have been no cases of 20 years, we are now moving toward the classification "chronic", [but] we're not there yet." (physician)

When questioned about PAH prognosis, the doctors did not have a clear, solid vision of PAH prognosis grounded in scientific evidence. Rather, they pointed out the complexity of the disease and the persistence of uncertainties:

"As for knowledge of the disease, detection of the severity of symptoms, and the strategy of treatment, we don't have much to go on." (pulmonologist)

"The natural history of PAH is not known. It is unclear when the symptoms occur... The symptoms are related to a decrease in cardiac output." (pulmonologist)

**PAH diagnosis**

The physicians noted that the period of diagnosis – estimated by some at "2 years" – is "long." They attributed this length of time to several factors: the non-specific nature of the symptoms; poor analysis of dyspnea by some primary care physicians; absence of the disease in diagnostic hypotheses of primary care physicians due to its rarity; and patients' delay in seeking medical advice, which retards diagnosis. Physicians disagreed on the impact on patients of this lengthy diagnosis. Some considered it a delay that affects the lives of patients, reduces their quality of life, and influences the prognosis for patient survival. Other physicians stressed the importance of the degradation rate of the disease; according to them, the causal link between the diagnostic period and worsening prognosis has not been proven.

**The impact of PAH on the daily lives of patients: an evaluation of their functional class**

In current medical practice, after diagnosis, PAH symptoms such as dyspnea, fatigue, chest pain, and discomfort lead physicians to administer a subjective medical evaluation. The NYHA Functional Classification System, modified by the World Health Organization assesses the degree of breathlessness experienced by patients [26]. This assessment is based on a clinical examination and on patients' reports on their capacity for exertion in their daily physical activities. Limitations are viewed as consequences of PAH symptoms and as indicators of the severity of the disease. Severity is a major prognostic factor, correlated with patients' life expectancy [27,28]. Physicians in this study noted imperfections with the NYHA instrument, describing it as ambiguous and subjective:

"The NYHA has an important predictive value... but remains incomplete or inadequate because we think it's a bit rough." (physician)

10 To respect the anonymity of respondents, only pulmonologists are identified; other specialists were too few to avoid revealing their identities.
Divergent approaches to assessing patients' functional class correspond to variations not only among physicians but also within physicians' own evaluation routines. Some physicians viewed NYHA as merely a way of classifying dyspnea, whereas others see it as a functional evaluation system:

"(When you say "NYHA", what does it mean for you?) It isn't dyspnea. It's the functional class. It's dyspnea, fatigue or pain, I think, and then... It's not the same for everyone, including colleagues who are super experts... and who pay more attention to dyspnea [when they assess functional class patients]. We all feel this problem." (pulmonologist)

"(What exactly does the NYHA scale evaluate?) The NYHA scale rates your current degree of dyspnea over the past two weeks, barring a pneumonia that may show up unexpectedly and change things." (physician)

The results showed that physicians' practices of patient assessment vary, as does their consideration of symptoms in the NYHA functional classification:

"(So far, you've just mentioned dyspnea...) Dyspnea is the symptom that interests us the most because it is generally the identifying sign, the first symptom." (physician)

Certain doctors used parameters in addition to those of the NYHA functional classification, integrating the results of medical examinations – specifically data from right heart catheterization, echocardiography, or walking tests. They also looked for other signs of right heart failure such as discomfort or syncope (included in the NYHA functional classification); they evaluate the progression rate of the disease; and they appraised their "overall" personal view of the patient:

"(Is questioning patients the only way to establish the difference between classes?) I also look at the walking tests. Overall, people who are in class II do around 400 meters, the people who are in class III do around 300 meters, and the people in class IV do around 200 meters. These are the averages we get when we study large cohort of patients. So if I have someone rated class II who only does 200 meters, I think there's a problem: either I interviewed them poorly, or they did their walking test half-heartedly. I take another look at things. I ask them whether pain affected their walking test and [I urge them] to reconsider the daily life examples they gave." (pulmonologist)

"You look to see if there are any immediate serious indicators – whether there are signs of right heart failure, edema, pain in the liver, syncope, chest pain on exertion, items that indicate severity. You identify that through questioning the patient. It's not complicated: you can conduct a clinical examination quite rapidly. If they don't volunteer it, you ask them 'Does your chest tighten up when you make an effort?'" (pulmonologist)

For this assessment, physicians did not always consider patients' perceptions relevant or sufficiently informative:

"Dyspnea is a medical word. We translate it as 'shortness of breath'. Is it discomfort, pressure, pain, panting, blockage...? Each person uses a different word depending on their culture, their origins, their feelings, the discomfort that they feel." (physician)

To overcome the shortcomings of the NYHA tool, some doctors created a fifth, intermediate class that allows a more precise evaluation of the functional condition of certain patients in order to monitor the progression of patients' health and treatment efficacy more accurately. Classes II and III were deemed too "broad" and do not indicate precisely enough patients' sensitivity to change. Other doctors nuance classes II, III or IV, by adding "low" or "high" or " + " to fine-tune their assessment. In some cases, this simply reflects an inability to objectively distinguish functional class II from III, or class III from IV.

**PAH treatment**

The doctors furnished a relatively paradoxical description of the therapeutic decision in PAH. Although they described a complex decision process, marked by scientific uncertainty, the NYHA system and international guidelines do provide a framework that makes therapeutic strategies systematic.

However, behind the apparent consensus concerning the NYHA functional classification, there is a relative heterogeneity in therapeutic strategy. The main difference is the degree of "aggressiveness" of the strategy adopted. Doctors had 2 different approaches: some advocated a "rapidly aggressive" approach, whereas the others favored a more conventional and gradual approach.

An ethical question nevertheless arises. There is a problem reconciling improving patients' quality of life and extending their life expectancy:

"Here you have several options, from simple to more aggressive and invasive... The real ambiguity is there. Suppose that a single treatment of 100 patients produces improvement for 20% of them, and thus the single treatment suffices. But it is not effective for 80% of them, and you may take 3–4 months to spot this. The question – which I have no answer to – is whether I should have immediately treated 100% of them more aggressively, since I have 80% that will not respond to single treatment, instead of taking 3–4 months in order to observe these 80 patients, knowing that by doing so, I might be over-treating 20% of the patients who needed a lighter treatment." (physician)
Patients’, relatives’ and practitioners’ views of pulmonary arterial hypertension: A qualitative study

Doctors try to find a balance between the risk of “over-treatment”, which may make patients addicted to a needlessly heavy and expensive treatment, and the risk of insufficiently treating the disease. For some doctors, the dilemma seemed to arise in the following terms: “Which do we want to do more, improve patients’ quality of life or extend their survival?”

**Physicians’ expectations regarding PAH**

Physicians were largely concerned about identifying and validating therapeutic strategies:

“The main challenge nowadays is the therapeutics. You can take the same patient, for example, and say that you absolutely must put him on oral bitherapy; another doctor will say, ‘No, he should be put on monotherapy.’ Or a patient that you think must absolutely be immediately given invasive treatment — there is another doctor who will say ‘No, we must first put him on oral bitherapy and then we’ll see.’” (physician)

They also stressed the need to review the NYHA functional classification by defining the indicators more precisely and possibly creating a fifth class:

“Between you and me, the ideal would be for the NYHA scale to have five classes.” (physician)

“The best thing to do would be to formalize a ‘3-weak’ and a ‘3-strong.’” (physician)

**Relatives’ views of PAH**

**PAH: a constructed reality**

Like patients with PAH, their relatives did not have a particular representation of PAH initially. It is an unknown disease, difficult to understand. Some relatives confused it with hypertension. This did not facilitate acceptance of the disease and the functional limitations that it causes; hypertension is viewed as a more common disease, not so “serious.”

**PAH diagnosis**

Throughout the patients’ process of “constructing” the first symptoms of PAH, their relatives participated, de-emphasizing or even denying the symptoms. They scoffed, questioned patients’ judgment, and suspected them of exaggerating their symptoms. Even though symptoms were visible, it did not mean that relatives systematically recognized and acknowledged patients’ limitations. However, as the symptoms intensified, family and friends revised their view of the patient’s complaints and did wind up telling them to see a doctor:

“The past few months, she couldn’t take the train — she had to lug her suitcase. . . We put it down to something else, but she was tired. She went to bed early, at 9:00 she was out, and it was practically impossible for her to get up the next day. It was always difficult. I remember one Monday, she was resting, she slept almost all the time, she didn’t move. At that point, we began to worry.” (relative 4)

“I remember one scene that shocked me. The children were young at the time. One day when he was angry with our son, he couldn’t muster any force. He didn’t have enough breath to get angry . . . Because it dragged on for years. For several years, it got worse, bit by bit.” (relative 1)

The announcement of the diagnosis disrupted the patients’ relations with family, friends, and spouse. Some reported a strengthening of ties when confronting PAH. Others, especially those in denial about the disease, stated that a distance was created, which caused problems when they were called on to assist with some or all of the activities that the patient could no longer do.

When relatives discussed the amount of time it took to diagnose the disease, they reported that patients tended to underestimate the time.

**The therapeutic management of patients: exclusion of relatives**

Relatives regretted not being included in patients’ evaluations; they indicated that patients minimized certain aspects of the disease, such as dyspnea, and its actual impact on their daily lives:

“I don’t remember when, but [my husband] spoke about [going up] three floors. It was recently, and I definitely noticed it. It was on the phone. But, I think I would have reacted in front of him, talking to the doctor. (You would have had the nerve to speak up to a doctor?) Oh yes! For a big thing like that, yes. But it’s true when he’s seeing the doctor, unless I make a point of intervening, I am rarely, very rarely asked my opinion.” (relative 1)

They expressed their dismay at particular risks associated with the disease, and at their lack of the information needed to manage problems such as fainting. Some would have liked to receive counseling to avoid expressing a negative view of the disease that impacted patients and their relationships.
Whereas they expected a treatment that would allow them and their ill relative "to live like before," they would have at least liked to minimize the side effects of treatment experienced by their relative with PAH, and facilitate medication administration in order not to disrupt activities outside the home. They also supported patients' wish for greater freedom of choice to accept or reject a caregiver who does not inspire confidence or to discontinue treatment protocols when they so desire. Relatives also wished for better management of pain. They expected clearer information about the mechanisms of drug action and about what signs constitute a "cure," fearing that the ill effects of a treatment might outweigh its benefit.

**Discussion**

This is the only available qualitative study that combines 3 complementary perspectives on PAH, those of the patients with PAH, their relatives, and the physicians specializing in PAH. It thus responds to limitations identified by other studies focused solely on the point of view of the patient with PAH [22]. Indeed, adopting a patient-centered approach cannot be limited to considering merely the patients' perspective. Sources of information must be triangulated, because the social reality studied – PAH in this study – emerges from the interaction between patients and their social environment. Relatives may voice different views of the disease, and their accounts can supplement the testimony of patients. Bringing together patients' and physicians' perspectives enables researchers to grasp the "signs" through which each side deciphers the disease – including the way they determine symptoms, going beyond strict clinical signs.

The results of this study showed certain discrepancies among those views. Patients with PAH, relatives, and physicians conveyed information that does not always concur, even when they addressed the same topics, such as symptoms, impact on daily life, and expectations.

PAH was thus shown to be a complex disease that was difficult to understand for patients and relatives, which physicians failed to explain satisfactorily. This difficulty can lead to confusion between PAH and systemic arterial hypertension.

The patients' semiology of the disease included more symptoms than those examined by the physicians. Their description of PAH impact on their lives conveyed a broader view than that of the doctors, which was mainly focused on functional aspects. The relatives' points of view appeared valuable because they provide a counterweight to the patients' evaluation of their condition; patients displayed a pattern of minimizing their symptoms in the results.

The list of symptoms identified by the patients also included more symptoms than identified in other studies. McDonough et al. [21] reported shortness of breath, dyspnea on exertion, coughing, chest pain, dizziness, fatigue, jaw pain, and forgetfulness. Our results do not include the latter but do additionally list edema, syncope, Raynaud's phenomenon, cyanosis, hemoptysis, palpitations, leg and liver pains, cramps and stiffness, insomnia, taste and olfactory disgust, weight variations, and an exacerbated sensitivity.

The results confirmed that the patient trajectory begins long before PAH diagnosis. They go through a progressive process of determination of their symptoms. We have observed a 4-step pattern leading from dismissal of their symptoms to the decision to seek medical advice. This pattern is globally consistent with Armstrong et al.'s results [22]. However, our results emphasized the social dimension of the construction of the symptoms, which are identified in relation to the patients' social environment, with relatives playing a role. Relatives do not always catalyze the decision to seek medical help; they may actually prevent identification of the symptoms. The decision to seek medical assistance is determined by the patients' failure to provide a satisfactory interpretation for the symptoms they experienced: patients decide to call for medical help when their symptoms no longer have any meaning for them other than that they are experiencing a health disorder.

As the study showed, this process was one part of the time needed to reach a diagnostic. However, the second part was the medical time needed once medical advice is requested. The existence of this diagnosis time was supported by other studies that qualified it as a "delay" to diagnosis [21]. In 2006, the results of the French national registry of PAH patients showed an average lapse of 27 months between the onset of the symptoms and a confirmed PAH diagnosis by right heart catheterization [2]. They also indicated that 75% of patients were diagnosed with stage III or IV according to NYHA classification. The issue then becomes whether the time required for diagnosis has a harmful effect on prognosis. Although the physicians interviewed did not agree on this point, they nonetheless agreed on the need for improving primary care dyspnea examination practices and for accelerating referral of patients to specialized centers.

The length, management, and accuracy of the diagnosis process are common issues in rare diseases. This point has not been well studied, but it has been mentioned for idiopathic pulmonary fibrosis [30-33] and pulmonary lymphangioleiomyomatosis [34]. However, these studies only point out the determinants related to organizing medical care (doctors' coordination, the multidisciplinary dimension of patient evaluation, referral to specialized centers, systematic application of guidelines, etc.) [30], without considering that the length of the diagnostic process also depends on patients and their social environment. Although all groups of interviewees were concerned by the impact of PAH, the patients in this study had a broader view, which included functional, financial, identity, and social dimensions, while physician were more focused on functional aspects. This perception gap may lead to differences in prioritizing therapeutic aims, in particular when treatment impacts patient
quality of life. This gap is reinforced by differences in the expectations of study patients, relatives, and physicians. Physicians' expectations focused more on identifying and validating therapeutic strategies, whereas patients and their relatives expected improvement in PAH diagnosis and the management of their treatment.

Furthermore, our study suggests that discussion of prognosis is difficult, pointing out a need for better doctor-patient and doctor-relatives communication, and for possible guidelines to help doctors announce prognosis to their patients without destroying all hope, as it has been already pointed out for patients with lung and colorectal cancer [35,36].

The study also points out that physicians have divergent approaches to assessing patients' NYHA functional class, which was also indicated by other studies [37]. This classification plays a major role in the choice of initial treatment, of assessment of the treatment benefits, and of therapeutic adjustment [38]. Our results lead us to question the way in which physicians interpret and use the NYHA functional classification and point out the need for greater standardization of these practices. The results also show that symptoms are "interpreted" through socio-cultural patterns that are partly determined by professional standards but also by individual views of quality of life, expectations, and perceived risks. The meaning of a symptom and the notion of effort also change throughout the patient therapeutic journey; an "ordinary activity" can become an effort at a different stage of this trajectory.

Study limitations

All patients with PAH interviewed were enrolled in the same medical department. Although the patients were not all seeing the same practitioner, their views of PAH are probably affected by ideas shared by professionals working in this department, the French National Center for Severe Pulmonary Hypertension. It is likely that their doctors had similar views on the disease, even though other medical departments may have other informational messages. Further research would be useful in order to see whether other patients' understanding of the disease includes aspects other than mere complexity.

Another limitation is the fact that mainly idiopathic PAH patients were considered. The patient sample included 12 patients with idiopathic PAH (the other 4 had heritable PAH), which has been shown to have a better prognosis than other forms of PAH especially associated with systemic sclerosis [39]. Further research would be necessary to assess whether patients with different forms of the disease have similar views on PAH.

Data saturation (i.e., the point where no new information emerges from the field) was reached in the interviews with patients and doctors. This was not the case with the relative sample, which was too small to provide a complete and deep overview of relatives' views. The interviews with relatives nevertheless provided a valuable counterpart to patients' narratives, highlighting in particular a potential discrepancy between what patients with PAH might report concerning their activities and their functional abilities, and the reality of their daily practices. Relatives also expressed their specific needs for information and therapeutic education in the interviews. Their views provided some preliminary exploratory data that further study would need to support.

The qualitative design of the study has provided extensive information on patient's views about their symptoms, the impacts of PAH on daily life, and their expectations. A quantification of these data would help in prioritizing areas of improvement in medical care.

Conclusions

Our results provide a unique and deep insight into patients', relatives, and physicians' views of PAH, and they suggest several potential improvements in patient management. More attention and time should be devoted to patients' complaints and preferences in order to better understand their priorities, including psychosocial aspects of their health such as quality of life. Current guidelines emphasize the importance of treat-to-target strategies in PAH. In a context in which few data are available to support the choice of adequate combination therapy, it might help to include patients' views, goals, and preferences in order to enhance treatment strategies and allow physicians to better tailor treatment to patients' needs. Moreover, our results clearly indicate that the assessment process for the functional impact of PAH should be re-examined to achieve a more uniform approach, and that patients' expectations regarding the effect of therapy may differ from those of physicians, as reported in other areas [36,40,41]. Further studies should be undertaken to devise a less ambiguous tool for patients' functional assessment. Our findings may be useful for improving therapeutic education for patients and their relatives, and also for enabling practitioners to better interpret dyspnea in patients and refer them for further medical assessment when appropriate. Finally, this qualitative study may also help develop patient-reported outcome measures with better content validity. It lays the groundwork for the development of new instruments to investigate the impact of PAH on patients' daily lives in terms of symptom assessment and functional impact.

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