The impact of pulmonary arterial hypertension (PAH) on the lives of patients and carers: results from an international survey
This large scale PAH Patient and Carer Survey was carried out to provide new insights into the wider impact of PAH on patients and carers beyond the clinical definition of the physical burden of the disease, which until now, has not been extensively researched. It explores four main areas: the physical and practical impact of PAH; the emotional impact; the financial impact; and information needs and provision; to better understand PAH patients’ and carers’ experience of living with the disease and to inform research and provision of more comprehensive care in the future.

The survey provides an international perspective, with respondents from five countries in Europe: France, Germany, Italy, Spain and the UK, all with different healthcare systems. For the first time, it explores the carer’s perspective as well as the impact of PAH on patients.*

Generally, the findings across the countries were consistent and reveal not only the major limitations that PAH has on the ability of patients to carry out everyday activities however small, such as household chores, shopping for food or taking a shower, but also the financial impact due to limitations on the ability of patients and carers to work. Last but not least, the results highlight the social isolation that the disease causes, especially among family and friends. The survey shows that a decline in patients’ functional class (FC) (a classification of severity of the condition, with FC I being the least decline in function and IV being the greatest) is associated with a greater impact on a wide range of factors beyond just physical activities, including relationships, the ability to work and to socialise, and emotional well-being.

Findings from the carers’ survey underline the need to include them as stakeholders in all aspects of the management of PAH. Most carers reported that caring for someone with PAH had had a very significant impact on their lives, and one in three had seen a reduction in their income, largely due to having to give up work or changing their working hours to take on a caring role.

As the steering committee of PAH specialists and representatives of patient organisations that led the development of the survey and this report, we hope that these important findings will help healthcare professionals and patient organisations to take account of the broader impact of PAH on patients and carers beyond the current definition of clinical symptoms and to provide care that meets these needs.

Acknowledgements: Our thanks to the PAH patients and carers who took part in the surveys and to the Associazione Ipertensione Polmonare Italiana Onlus (AIPI), Associazione Malati di Ipertensione Polmonare (AMIP O.N.L.U.S.), Asociación Nacional de Hipertensión Pulmonar (ANHP), Association des Sclérodermiques de France (ASF), Fundación Contra la Hipertensión Pulmonar (FCHP), HTAP France, PHA UK, Pulmonale hypertonie e.v. who gave valuable input into the planning and recruitment of this research.

* A selection of questions asked during the survey can be found throughout this report.
The impact of pulmonary arterial hypertension (PAH) on the lives of patients and carers: results from an international survey

September 2012
Sponsorship: The survey and report have been developed under the auspices of a steering committee of PAH specialists and representatives of PAH patients’ organisations supported by Actelion Pharmaceuticals Ltd, Switzerland. The market research agencies involved in the surveys were commissioned by Actelion.

EXECUTIVE SUMMARY

BACKGROUND
Pulmonary arterial hypertension (PAH) is a rare condition but one which has a major impact on morbidity and mortality and currently there is no cure. There has previously been relatively limited research into the broader impact of PAH on the lives of patients and carers beyond the current definition of clinical symptoms, so an international self-reported survey using market research methodologies was carried out to explore this. A multidisciplinary steering committee of PAH specialists, including respiratory physicians, intensists, rheumatologists, PAH specialist nurses and patient group representatives, led the development of the survey content and analysis of results to ensure medical and practical relevance.

METHODS
A total of 455 PAH patients (n=326) and carers (n=129) from five European countries (France, Germany, Italy, Spain and the UK) replied to questionnaires asking about four main areas: the physical and practical impact of PAH; the emotional impact; the social impact; and information needs and provision. This quantitative data was informed by qualitative interviews with 25 PAH patients and 15 carers to provide information on key themes to further explore in the online/postal questionnaires.

RESULTS
A total of 455 PAH patients (n=326) and carers (n=129) from five European countries (France, Germany, Italy, Spain and the UK) replied to questionnaires asking about four main areas: the physical and practical impact of PAH; the emotional impact; the social impact; and information needs and provision. This quantitative data was informed by qualitative interviews with 25 PAH patients and 15 carers to provide information on key themes to further explore in the online/postal questionnaires.

KEY FINDINGS
More than half (56%) of patients and a similar proportion of carers (57%) said PAH had a ‘significant’ impact on their daily life, which was associated with decline in functional class (FC). Employment/work and income emerged as areas of particular concern for both patients and carers: 85% of patients and 29% of carers said their work was affected by PAH. Nearly three-quarters (73%) of patients said their household income* was reduced by PAH, leaving most families with below average household incomes. Many patients reported that their sex lives were severely affected; as did carers who were partners of PAH patients.

Over half (55%) of patients and one-third of carers reported feeling isolated, with a major reason being a lack of understanding of the disease among family and friends, as well as among the general public. The emotional impact on patients of having PAH included feelings of frustration (33%) had felt this in the last month); feeling misunderstood; little pleasure in activities; anger and low self-esteem.

Nearly 25% of patients felt stressed whilst caring for their relative and trying to meet their other responsibilities, which increased with decline in the patient’s FC. Despite the major social, financial and emotional impact of PAH, patients and carers said they had been given little information on these aspects by healthcare professionals (HCPs), with patient organisations emerging as major sources of information and support.

RECOMMENDATIONS
Results from this first large international survey to explore the broader impact of PAH show that patients with PAH should be offered a comprehensive standard of care to assess and improve their emotional and social well-being, as well as their physical symptoms, recognising that needs increase with decline in FC. Optimal PAH management requires a multidisciplinary approach, including physicians, nurses, social workers, psychologists and patient organisations along with carers. The needs of carers are currently underestimated and information needs and provision. This quantitative data was informed by qualitative interviews with 25 PAH patients and 15 carers to provide information on key themes to further explore in the online/postal questionnaires.

PAH: histopathological features

WHAT IS PAH?
Pulmonary arterial hypertension (PAH) is a progressive disease caused by narrowing and tightening (constriction) of the pulmonary arteries, which connect the right side of the heart to the lungs. By definition, PAH is characterised by an increase in mean pulmonary arterial pressure (PAP) to at least 25 mmHg at rest, and a mean pulmonary capillary wedge pressure (PCWP) of <15 mmHg.1, 2

As PAH develops, blood flow through the pulmonary arteries is restricted and the right side of the heart becomes enlarged due to the increased strain of pumping blood through the lungs. It is this strain on the heart and the decrease in blood to the left heart and systemic circulation through the lungs that leads to the common symptoms of PAH, such as breathlessness, fatigue, weakness, angina, syncope, and abdominal distension.2

Although there is no cure, treatment is aimed at improving symptoms, exercise tolerance, long-term outcomes and quality of life. Until the mid-1990s there were limited treatment options for patients and PAH was associated with poor prognosis.

WHAT CAUSES PAH?
Although PAH is a rare disease, with an overall estimated prevalence of 15-50 cases per million, the prevalence of PAH is substantially higher in certain at-risk groups, including patients with human immunodeficiency virus (HIV) infection and those with systemic sclerosis.

WHAT IS THE IMPACT OF PAH ON PATIENTS?
Changes to the pulmonary vasculature lead to the typical symptoms of PAH, which include: breathlessness (dyspnoea), particularly during physical activity; fatigue; dizziness; syncope, also on physical activity; peripheral oedema and chest pain, again, particularly during physical activity.2, 3 These symptoms can severely impact a patient’s ability to carry out normal daily activities.

As the disease progresses, some patients may experience constant dyspnoea and fatigue so that even simple tasks, such as getting dressed and walking short distances, become difficult.

Illustration 1. PAH: histopathological features

WHAT MAKES THE IMPACT OF PAH ON PATIENTS?

The clinical severity of PAH is classified using a system that grades PAH severity according to the functional status of the patient, linking symptoms with activity limitations.2 It allows clinicians to quickly and accurately assess disease progression and prognosis, as well as the need for specific treatment regimens, irrespective of the underlying aetiology of PAH (Table 1).

*Participants were not asked for net or gross income so results assume after tax.
This research was set out to explore four main areas: physical and practical impact of PAH; emotional impact; social impact; and information needs and provision; using market research methodologies (Table 2).

Table 2: Schematic representation of questionnaire

Table 3: Background information on PAH: Patients self-reported awareness, understanding & relevance of FC

It was carried out in two phases:

1. **Phase 1** was a qualitative survey based on one-to-one interviews with trained market research interviewers and 25 patients and 15 carers across five European countries (France, Germany, Italy, Spain and the UK), designed to identify themes to explore further.*

2. **Phase 2** was a quantitative survey using opinion-based online or postal questionnaires in 326 patients (average age 52 years; 74% female; 5% FC I, 20% FC II, 34% FC III, 11% FC IV (self-reported FC)) (Table 3). A further survey was carried out in 129 carers (average age 52 years; 56% female; 3% caring for a patient with FC I, 31% FC II, 43% FC III, 23% FC IV (self-reported FC)) from the same five countries. The carers’ sample was not cross-matched or paired to the patients’, with a larger proportion of carers caring for patients of FC III and IV (66%) than reported in the patients’ survey (43%).

Responders were recruited mainly by local PAH patients’ organisations, apart from Italy, where participants were also recruited via advertisements. The sample size differed between countries due to feasibility of recruitment, therefore, statistical weighting was applied to the survey data to ensure the sample had the same weight/importance in the final sample as a whole.

The surveys were completely confidential and participants were guaranteed anonymity except for any disclosure to which they specifically consented. A multidisciplinary steering committee of PAH specialists, including respiratory physicians, intensivists, rheumatologists, PAH specialist nurses and patient group representatives, led the development of the survey content and analysis of results to ensure medical and practical relevance. The group developed a call to action setting out recommendations that aimed to meet the needs of PAH patients and carers identified by the survey findings.

1. **KEY RESULTS: PHYSICAL AND PRACTICAL IMPACT OF PAH ON PATIENTS**

   **In the last month how often have you experienced the following?**

   **RESTRICTION OF PHYSICAL AND EVERYDAY ACTIVITIES**

   Patients reported their physical activities were severely restricted due to PAH symptoms, with the most common being non-specific symptoms, including fatigue (59% of patients experienced this often or very often in the last month), breathlessness (56%), lack of energy (51%) and loss of libido/interest in sex (44%) (Chart 1).

   **On a scale from 1 to 10, how would you describe the impact of your PAH on your daily life?**

   Patients were frequently unable to perform everyday or ‘normal’ domestic tasks, however small, such as household chores and shopping for food; 56% of patients said that PAH had a ‘very significant’ impact on their daily life, while a further 23% said it had a ‘moderate impact’.

   A decline in patients’ FC was associated with an impact on daily life: 36% of patients in FC II were ‘very significantly impacted’ by PAH, increasing to 78% of patients in FC III.

   The key areas of life impacted by PAH were:

   - **Physical activities** – overall, 97% of participants taking part in the survey found their ability to travel or go on holiday impaired.
   - Employment/work – overall, 85% of patients found their employment/work was affected by their condition.
   - **Travel and social opportunities** – overall, 77% of PAH patients reported their ability to travel or go on holiday was impaired; 55% said leisure and cultural activities were

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Table 1. WHO functional classification of pulmonary hypertension

<table>
<thead>
<tr>
<th>FUNCTIONAL CLASS</th>
<th>SYMPTOMATIC PROFILE</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Patients with pulmonary hypertension but without limitation of physical activity. Ordinary physical activity does not cause dyspnoea or fatigue, chest pain, or near syncope</td>
</tr>
<tr>
<td>II</td>
<td>Patients with pulmonary hypertension resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnoea or fatigue, chest pain, or near syncope</td>
</tr>
<tr>
<td>III</td>
<td>Patients with pulmonary hypertension resulting in marked limitation of physical activity. They are uncomfortable at rest. Less than ordinary activity causes undue dyspnoea or fatigue, chest pain, or near syncope</td>
</tr>
<tr>
<td>IV</td>
<td>Patients with pulmonary hypertension with an inability to carry out any physical activity without symptoms. These patients manifest signs of right heart failure. Dyspnoea and fatigue may even be present at rest. Discomfort is increased by any physical activity</td>
</tr>
</tbody>
</table>

* Percentages in brackets are percentages of the total sample.

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**Table 2. Schematic representation of questionnaire**

Test: 59 questions, which were mostly with multiple choice possible

**Table 3. Background information on PAH: Patients self-reported awareness, understanding & relevance of FC**

My sister’s (patient) main objective is to get home to her house, she’s afraid of the hospital. Yesterday on the motorway I did 130kph because all she wanted was to get home. Carer

She manages to cook at times, but doesn’t go over 10kph. Patient

I used to sleep 5-6 hours a night and feel well. Now I sleep 8 and I’m tired all the time. Carer

I would describe PAH as being like going in a car which won’t go over 10mph. Patient

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**Chart 1. How patients assess the impact of PAH on their daily lives**
affected and 53% reported an impact on playing with their children or grandchildren.

- Dealing with domestic work/household chores – overall, 83% of patients taking part in the survey found it difficult to deal with domestic work and household chores; 72% found that going on errands such as shopping for food was affected by their condition.
- Relationship issues – being fully intimate with a spouse/partner was reported as a key area affected by PAH in 63% of patients taking part in the survey; overall, 58% said they found taking on the role of spouse/partner difficult.

How would you indicate your ability to perform or carry out the following activities compared to your ability prior to your diagnosis?

Chart 2. Patients’ indication of ability to perform activities post diagnosis

<table>
<thead>
<tr>
<th>Activity</th>
<th>No impact at all</th>
<th>Some difficulty</th>
<th>Can do it, no matter how much I want to</th>
<th>Can do it, but with a lot of difficulty and may need help</th>
<th>Cannot do it anymore and need help</th>
</tr>
</thead>
<tbody>
<tr>
<td>Taking a shower</td>
<td>3%</td>
<td>25%</td>
<td>38%</td>
<td>16%</td>
<td>1%</td>
</tr>
<tr>
<td>Cooking</td>
<td>3%</td>
<td>16%</td>
<td>25%</td>
<td>43%</td>
<td>9%</td>
</tr>
<tr>
<td>Doing domestic chores</td>
<td>3%</td>
<td>11%</td>
<td>25%</td>
<td>43%</td>
<td>9%</td>
</tr>
<tr>
<td>Taking on the role of spouse/partner</td>
<td>37%</td>
<td>40%</td>
<td>32%</td>
<td>12%</td>
<td>1%</td>
</tr>
<tr>
<td>Being fully intimate with my spouse</td>
<td>37%</td>
<td>40%</td>
<td>32%</td>
<td>12%</td>
<td>1%</td>
</tr>
<tr>
<td>Climbing a flight of stairs</td>
<td>77%</td>
<td>21%</td>
<td>9%</td>
<td>2%</td>
<td>1%</td>
</tr>
<tr>
<td>Walking a short distance</td>
<td>37%</td>
<td>43%</td>
<td>16%</td>
<td>25%</td>
<td>9%</td>
</tr>
<tr>
<td>Visiting relatives/friends</td>
<td>57%</td>
<td>39%</td>
<td>5%</td>
<td>13%</td>
<td>3%</td>
</tr>
<tr>
<td>Having a telephone conversation</td>
<td>81%</td>
<td>11%</td>
<td>3%</td>
<td>1%</td>
<td>1%</td>
</tr>
<tr>
<td>Visiting relatives / friends</td>
<td>57%</td>
<td>43%</td>
<td>3%</td>
<td>13%</td>
<td>3%</td>
</tr>
<tr>
<td>Playing with my children or ...</td>
<td>81%</td>
<td>2%</td>
<td>9%</td>
<td>2%</td>
<td>1%</td>
</tr>
</tbody>
</table>
| Employment/work was the activity that patients were most concerned or frustrated about in terms of the impact of PAH on their activities, and this had major implications for household income.

Have you had to stop working since being diagnosed with PAH?

Overall, 85% of patients taking part in the survey said their employment/work was affected by PAH, with nearly half (48%) saying they were unable to work anymore or needed help to do so. This increased with decline in FC: nearly all (93%) of patients with FC IV had stopped work or needed help to work as a result of their condition, compared to 66% of those with FC III, 29% of those with FC II and 5% of those with FC I.

The reduction in ability to work had a major impact on household income. Nearly three-quarters (73%) of patients who had to stop or change their work due to PAH said this had affected their overall household income*, leaving most families with below average incomes. Just over one in six (16%) of these patients said their household income had more than halved.

Could you please estimate the impact* it had on the overall income of your household?

*Question asked of respondents who reported an impact on the overall income

The financial impact on overall household income due to having to change working activity increased with decline in patients’ FC. From 32% of patients with FC I reporting an impact on household income, to 67% of those with FC II, 79% of those with FC III and nearly all (95%) of patients with FC IV.

*Lack of understanding of my disease by my children or grandchildren

If you have experienced loss in libido/interest in sex since your diagnosis, what has caused it?

Several factors may cause a loss of libido in PAH patients. The survey results showed that 30% of those reporting loss of libido thought this was caused by low self-esteem/body image, 23% were physically not able because of their PAH, 14% had one or more serious conditions in addition in PAH, 12% were afraid of becoming more ill and 6% blamed fear of pregnancy.

**PHYSICAL AND PRACTICAL IMPACT OF PAH ON PATIENTS**

The research highlighted the nature and degree of the physical restrictions on patients with PAH. There was particularly marked decline in ability to work, exercise and travel as the FC declined. In addition there was a knock-on effect of PAH on household income with a majority being adversely affected. The physical impact of PAH extended to loss of intimacy and lack of interest in sex, which grew worse as FC declined.

2.0 KEY RESULTS: SOCIAL IMPACT OF PAH ON PATIENTS

Qualitative interviews revealed that the patient’s world narrowed as result of being confined to the home. This was manifested as the loss of many of the social roles that patients had once held, including those as a colleague, friend, and team-mate. Patients’ need for support had strengthened many family relationships, but also had some negative consequences. Husbands found their social identity was affected by no longer being able to carry out activities they saw as part of their role, including home maintenance and playing football with their children. Wives felt their social identity was reduced by not being able to perform maternal tasks with their children and household.

How would you describe the overall awareness of PAH?

Respondents felt there was a lack of knowledge and awareness of PAH among family, friends and the general public. This leads to little empathy being shown towards patients.

**They just think we’re lazy. They don’t understand I can’t do it, no matter how much I want to. Patient**

**They just think we’re lazy. They don’t understand I can’t do it, no matter how much I want to. Patient**

**Nobody understands my suffering and they don’t want to listen. Everyone, doctors especially, even my daughter. Patient**
Most patients surveyed (88%) considered that public awareness of PAH was non-existent or very low. This low level of awareness impacted on overall well-being in more than half (57%) of patients, particularly in young women, patients and men in more severe cases (FC IV, 79%). What is more concerning is that patients claim the level of information and awareness among their family and friends was low, with almost half of carers (40%) not understanding the concept of PAH, which impacted their recognition of patients’ limitations and the goals of clinical care.

On a scale from 1 to 10, how has your role/status in the following changed since your PAH diagnosis?

Independence had been significantly reduced as a result of having PAH; 49% considered their role in society had changed and 42% felt there had been a change in their personal relationship with their partner or spouse.

In most cases (85%), the patient’s main carer was a member of their family – mainly their spouse or partner (61%). Just over two-thirds (68%) of patients said their carer had had a positive impact on their quality of life. This perceived contribution to quality of life seemed to increase with disease severity, from 40% in FC I to 53% in FC IV.

SOCIAL IMPACT OF PAH ON PATIENTS

The research highlighted the significance of the carer. The majority of patients recognised the positive contribution of their carer towards their quality of life. The claim of low public awareness of PAH is perhaps unsurprising given the rarity of the condition. However, it was notable that patients had poor awareness among their colleagues, friends and even close family members had a significant impact on the patient, making them feel isolated by their PAH.

5.6 KEY RESULTS: EMOTIONAL IMPACT ON PATIENTS

The qualitative interviews demonstrated a disconnect between what patients were saying their lives were like and their underlying feelings. The emotional toll of PAH was all encompassing, however, patients placed a stronger focus on their unmet physiological needs, for example breathing difficulties. Their full feelings were often not voiced, although signs of them were unconsciously leaked, with patients often putting on a ‘brave public face’ even to carers and families. The huge physical, practical and social changes that PAH imposed on their lives resulted in a wide range of negative emotions, including fear, guilt, loss, worry, stress, loss of libido, feelings of sadness, loneliness, frustration, isolation and depression, which were interlinked.

The main emotions experienced often or very often by PAH patients taking part in the survey were: frustration (35% had felt often or very often in the previous month, which was the period asked about); feeling misunderstood (26%); taking little pleasure in activities they used to enjoy (26%); feeling angry (24%); low self-esteem (22%); feeling worthless (22%); and feeling isolated (21%) (Chart 4). Other emotions included: feeling low in the morning, hopelessness, fear, low energy, and others.

Who is providing you with emotional support about living with PAH?

Unsurprisingly, it appears that family and carers are the greatest source of support: 42% of patients gained the most effective emotional support from their family and 29% from their carer. Only 14% of patients cited their specialist as providing ‘the most useful’ emotional support for living with PAH. Psychologist support appeared to be very low, with only 5% of patients saying they received this type of support despite the range of psychological symptoms they reported.

Where are you likely to go and look for information about PAH?

In the qualitative interviews, patients reported that they had been given very little information on the emotional impact of PAH by MCPs and what they had been told was presented verbally and forgotten due to the shock of the diagnosis. Very little information on the emotional impact of PAH seemed to have been given, with specialists focusing on physical symptoms, indicating scope for improved provision of comprehensive written materials on all aspects of the condition. PAH specialists about the role of patient organisations and the importance of working with them collaboratively.
The impact of PAH on carers and patients daily lives (as reported by carers)

The qualitative interviews showed that carers take on a ‘fighter’ and ‘organiser’ role, carrying out physical tasks and chores to compensate for patients’ inability to perform these themselves. This leaves carers physically drained, which is further compounded when carers are older and in ill health themselves. Interviews also highlighted the range of information that carers needed to help with their day-to-day caring role.

On a scale from 1 to 10, how would you assess the impact of your relative’s PAH on HIS/HER daily life?

Chart 6. The impact of PAH on carers and patients daily lives (as reported by carers)

Carers rated the overall impact of their relatives’ PAH on their daily lives as very high: 57% rated it between 8-10 on a 10-point impact scale (Chart 6). A higher proportion of carers of patients of FC III and IV reported a very high impact on their lives (53% and 88%, respectively) than those caring for a patient with PAH FC II (41%). The impact was also greater for carers aged 60 years and older (74% in this age group rating the impact as very high).

Would you please indicate all the changes your relative’s PAH has had on YOUR daily life?

PAH led to many changes in carers’ daily lives, with extra tasks and responsibilities resulting in exhaustion. Over half (51%) were more involved in household tasks, while 46% more likely to do go on errands and 45% were more involved in tasks that their relative used to manage (Chart 7). This had a major impact on carers’ lives: 45% felt exhausted more frequently, due to their extra responsibilities, and the same proportion had to schedule family life around the patient’s needs. One-third of carers said they needed to be organised all the time, requiring extra planning.

Phisical and Practical Impact of PAH on Carers

Patients report that little information is given at diagnosis and they are left with unanswered questions. Many look to the patient support organisations for their information and emotional needs to be met but many patients are not directed to this important source of support by their HCPs. Patients express interest in many aspects of PAH and its consequences, wanting some of those at time of diagnosis and others ‘fed through’ later (avoiding them becoming overwhelmed).

Table 4. Most suitable time for patients to receive topical information

At diagnosis seems to be the preferred time to receive information on certain topics (euphemistically referred to as diagnosis and treatment), while other topics become more important subsequently.

1.8 KEY RESULTS: PHYSICAL AND PRACTICAL IMPACT ON CARERS

The research highlighted the significant impact caring for someone with PAH has on the carer themselves. Being a carer leads to many changes in the carers own life as they need to help more with extra chores and always have to forward plan, resulting in them often feeling exhausted. Carers’ ability to work is impacted, which has a major effect on their income. Their ability to work declines, as the FC of the person they care for declines.

Carers who are partners of PAH patients also report a decrease in sexual relationships.

1.1 IMPACT ON EMPLOYMENT WORK

Nearly one-third (29%) of carers reported stopping work or changing their working conditions as a result of caring for a person with PAH, with 13% stopping work completely. Having to reduce or give up work as a result of caring for someone living with PAH had a major impact on carers’ incomes, with more than one-third (35%) of carers reporting a reduction in their income.

People caring for someone with PAH FC IV were far more likely to have given up work than someone caring for a person with FC II (20% vs. 6%), due to the extra tasks and responsibilities required to care for a person less able to do things for themselves. This then has a significant impact on carers’ financial circumstances.

One of the first things (it) said was I could live for 2.5 years with PAH - not pleased to read this straight after diagnosis. Patient
The time spent caring for someone with PAH increased as FC declined (FC II 49%, FC III 62%, FC IV 70%).

**DO YOU feel isolated/excluded at times as a result of caring for someone with PAH?**

Almost one-third (30%) of carers felt isolated/excluded as a result of caring. Of these, 61% found this had a very significant impact on their daily life. Feelings of isolation were greatest among carers who looked after patients with IPAH (38%) and in carers who had to stop working or change their working conditions (54%) (Chart 8).

On a scale from 1 to 10, what impact does feeling isolated/excluded have on your daily life?

Chart 8. Feelings and impact of PAH on carers

<table>
<thead>
<tr>
<th>Impact of isolation on daily life</th>
<th>Very significant impact</th>
<th>Significant impact</th>
<th>Moderate impact</th>
<th>Slight impact</th>
<th>No impact at all</th>
</tr>
</thead>
<tbody>
<tr>
<td>Feel isolated</td>
<td>70%</td>
<td>30%</td>
<td>31%</td>
<td>31%</td>
<td>31%</td>
</tr>
<tr>
<td>Average 6.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

I no longer have hobbies as it is not feasible with being a carer... It takes up all my time. It takes up your whole day caring for the patient. Carer

Lack of visibility of PAH and lack of understanding by family and friends and the wider community were major causes of carers’ feelings of isolation.

**SOCIAL IMPACT OF PAH ON CARERS**

The research highlighted that caring for someone with PAH can have a significant impact on the carers own social life. Carers are restricted socially as they spend a high proportion of their time caring, which can result in feelings of isolation.

The time spent caring increases as the functional class of the patient declines. The lack of visibility of the disease and lack of understanding by friends and family may contribute to these feelings of isolation.

5.8 KEY RESULTS: EMOTIONAL IMPACT ON CARERS

The in-depth interviews suggested that carers tend to prioritise the patient’s emotions above their own. That there was a strong sense of underlying frustration and carers appear to have strong emotional needs which are currently not being met.

In the survey, two-thirds of carers (66%) reported being concerned for the future for their relative often or very often. Nearly one-quarter (22%) of carers said they felt stressed caring for their relative and trying to meet their other responsibilities. This increased with decline in the patient’s FC (12% for FC II to 29% for carers of people of FC IV) (Chart 9).

Chart 9. Carers feelings when caring for a patient with PAH

<table>
<thead>
<tr>
<th>Impact</th>
<th>Carers commonly feel afraid for the future, feeling they should be doing more for the patient</th>
<th>Very</th>
<th>Slightly</th>
<th>Moderate</th>
<th>None</th>
</tr>
</thead>
<tbody>
<tr>
<td>I feel afraid about what the future holds for my relative (126)</td>
<td>12%</td>
<td>22%</td>
<td>31%</td>
<td>35%</td>
<td></td>
</tr>
<tr>
<td>I feel stressed between caring for my relative and trying to meet other responsibilities for my family or work (124)</td>
<td>20%</td>
<td>29%</td>
<td>33%</td>
<td>25%</td>
<td></td>
</tr>
<tr>
<td>I think I should be doing more for my relative (124)</td>
<td>29%</td>
<td>25%</td>
<td>34%</td>
<td>10%</td>
<td></td>
</tr>
<tr>
<td>Increases with FC: 12% of FC II; 27% of FC III; 29% of FC IV</td>
<td>51%</td>
<td>14%</td>
<td>12%</td>
<td>12%</td>
<td></td>
</tr>
<tr>
<td>I feel I have no choice but to look after my relative (121)</td>
<td>27%</td>
<td>23%</td>
<td>15%</td>
<td>17%</td>
<td></td>
</tr>
<tr>
<td>I feel I could do a better job in caring for my relative (122)</td>
<td>20%</td>
<td>25%</td>
<td>33%</td>
<td>17%</td>
<td></td>
</tr>
<tr>
<td>I feel uncertain about what to do for my relative (121)</td>
<td>15%</td>
<td>18%</td>
<td>12%</td>
<td>22%</td>
<td></td>
</tr>
<tr>
<td>I feel that my relative is overly dependent on me (123)</td>
<td>30%</td>
<td>16%</td>
<td>20%</td>
<td>13%</td>
<td></td>
</tr>
<tr>
<td>I feel that caring for my relative gives me less time to spend with my children or grandchildren (PARENTS/GRANDPARENTS only)</td>
<td>54%</td>
<td>21%</td>
<td>21%</td>
<td>15%</td>
<td></td>
</tr>
<tr>
<td>I feel that my health has suffered because I am caring for my relative (124)</td>
<td>55%</td>
<td>16%</td>
<td>15%</td>
<td>13%</td>
<td></td>
</tr>
</tbody>
</table>

Lack of visibility of PAH and lack of understanding by family and friends and the wider community were major causes of carers’ feelings of isolation.

**Lack of visibility of PAH and lack of understanding by family and friends and the wider community were major causes of carers’ feelings of isolation.**

The following statements reflect how people sometimes feel when they are taking care of another person. Please indicate how often, if ever, YOU feel that way.

<table>
<thead>
<tr>
<th>Statement</th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Often</th>
<th>VERY OFTEN (rarely/never)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Life with someone with PAH is like not knowing what to expect, I don’t know how it will develop. As a parent, not seeing him grow up and develop.</td>
<td>34%</td>
<td>29%</td>
<td>21%</td>
<td>15%</td>
<td>1%</td>
</tr>
<tr>
<td>I feel cross, frustrated and frightened for my future. Unless he has a lung transplant, he’s not going to be here this time next year, it’s not what we planned or wanted but we can’t go back.</td>
<td>14%</td>
<td>35%</td>
<td>31%</td>
<td>21%</td>
<td>9%</td>
</tr>
</tbody>
</table>
| Being a carer can have a positive impact, with 92% of carers feeling they are contributing to the patient’s quality of life. Nearly two-thirds (62%) considered that caring had brought them closer as a couple or family. Over one in three (34%) found this had a very significant impact on their daily life. Feelings of isolation were greatest among carers who had to stop working or change their working conditions (54%) (Chart 8).

Carers are restricted socially as they spend a high proportion of their time caring, which can result in feelings of isolation.

Nearly two-thirds (62%) considered that caring had brought them closer as a couple or family. Over one in three (34%) said they did not find fulfilment in being a carer, underlying the challenge in the change in role they have to cope with when they become a carer, which can be particularly obvious when they have to give up work.

The research has highlighted that carers put the feelings of the person they care for first, but that they themselves can experience feelings of stress and frustration and they often fear what the future will hold as it is unknown.

**EMOTIONAL IMPACT ON CARERS**

At diagnosis, most carers claim to receive little/no written information about PAH from HCPs which, due to the initial shock of diagnosis, leads to many unanswered questions. There was a particular lack of information about emotional aspects of PAH. Carers are heavily involved in attending patient appointments/groups and have a real need to find out additional information from a variety of sources about PAH.

Interviews found that carers are more pro-active than patients in sourcing information, more actively looking up additional information from a variety of sources e.g. internet, reading and DVDs and they filter information to patients based on how appropriate it is. Patient organisations are the most important source of information for carers but could provide more information about social support/financial assistance.

**4.8 KEY RESULTS: CARERS’ INFORMATION NEEDS**

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**What type of information on PAH do you feel YOU need or wish you received?**

Carers were keen to receive regular updates on their relative, detailed drug information and information on the disease itself from HCPs, with 87% looking to receive a regular update on the health status of their relative and potential progression of their PAH, alongside detailed information on drugs used in PAH therapy. More than one in five (23%) carers expect information about the financial and administrative consequences of PAH from HCPs, suggesting a need for further HCP education around wider provision of information on the impact of PAH on carers.

The information requested by carers was very similar to that wanted by patients, but carers were more interested in information on specialists and doctors involved in PAH care; the emotional consequences of having the condition; follow-ups including schedules and purpose; and financial consequences.

**PAH INFORMATION NEEDS: THE IMPACT ON CARERS**

The research has highlighted that carers actively seek information from a variety of sources and filter what information they pass on to the patient. Carers request similar information to patients but they express an interest in receiving information on specialists and doctors involved in PAH care, social support, financial support and emotional consequences of having the condition.

**Do YOU currently belong to any patient association/support group?**

Over two-thirds (68%) of carers had joined a patient organisation; one in four reported being active members. Leading reasons for joining patient organisations were wanting to know all about PAH (56%), and wanting to hear about and meet other patients and carers and how they were coping (48%).
The survey showed that employment and work was the activity that PAH patients across Europe were most concerned about in terms of the impact of PAH, and this had major implications for household incomes. Overall, 85% of patients said their employment/work was affected by PAH, with nearly 1/3 saying they were unable to work anymore or needed help to do so because of their condition. Capacity to work was also affected by PAH in nearly 1 in 5 patients taking part in the survey having stopped work or changed their working conditions as a result of caring for a person with PAH, and 13% giving up work completely. Nearly three-quarters of patients said their household income was reduced because of PAH, giving up work completely. Nearly three-quarters of patients and carers taking part in the survey saying that the disease burden of PAH increases significantly with a decline in FC. Unsurprisingly, a decline in FC was associated with a greater impact on daily life for the patients themselves and for carers.

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The results of this international survey have major implications for the scope of care provided to patients with PAH and underline the need to include carers as important stakeholders. They show that PAH guidelines need updating to reflect the multidimensional care necessary to address patients’ and carers’ emotional and social needs, as well as their physical needs. This care would best be provided by a multidisciplinary team of HCPs and other specialists, including physicians, nurses, social workers, psychologists and patient organisations.

Provision of clinical care should be adapted to meet the needs of the individual PAH patient, including consideration of their physical, social and emotional needs, and recognising that the disease burden of PAH increases significantly with a decline in FC. The important role of the carer should also be given due consideration.

There is a major need for improving the provision of all aspects of PAH. Patient organisations have an important role in this, as well as in providing support and networking for PAH patients and their carers. It is important that HCPs continue to refer patients and carers to patient groups so both can benefit from their help. Patient organisations and HCPs can work together to improve the provision of information on PAH and to lobby effectively for improved resources and support, and should be better supported in this. The ultimate aim should be to improve all aspects of the lives of patients with PAH and their carers – physical, social, emotional and emotional – so they can enjoy the best outcomes and quality of life that are achievable.

This international survey gives unprecedented insights into the wider impact of PAH on the lives of patients and carers beyond the physical burden of the disease and reveals the negative effects on their daily lives and activities and on their emotional and social well-being. Results show the huge changes that people have experienced, with many patients and carers describing striking changes in their lives.

The qualitative interviews uncovered a massive disconnection between what patients said their lives were like and their true feelings. The huge physical, practical and social changes that PAH imposed on their lives resulted in a wide range of negative emotions, including fear, guilt, loss, worry, feelings of uselessness and frustration. Only one in twenty patients said they received support from a psychologist, demonstrating the scope for improving provision of this type of help. Carers felt concerned for the future for their relative and nearly one in four felt stressed by juggling caring with their other responsibilities.

The PAH patients and carers surveyed felt there were gaps in the information they were given about the condition, with a particular lack of information about emotional aspects of coping with PAH and financial and administrative implications. Patient organisations emerged as playing a leading role in providing information and support for both patients and carers. Survey participants wanted to receive information about PAH and its treatment at the time of diagnosis but many said they would like to get information on further aspects at a later stage.
CALL TO ACTION FOR PATIENTS

Living with PAH is difficult and its impact on quality of life is far-reaching. It is imperative that patients are educated on and aware that the effects of PAH go beyond those of the clinically defined symptoms given at diagnosis and can have implications on practical, social, emotional and financial aspects of life.

1. Patients should expect access to the right information at the right time to help them live with PAH
   - Accurate and easily understandable information should be available within the specialist PAH hospital/clinic setting over a much longer period post diagnosis
   - Patients should be provided with support, which goes beyond that currently provided in clinical practice, to help patients to better explain their disease and its impact to others

2. Patients should expect access to a service with increased awareness of non-physical effects of PAH amongst its HCPs
   - Patients should feel confident in asking HCPs for advice and support beyond their physical care needs (including advice on social, emotional and financial aspects)
   - Further information on FC and the impact of FC severity on quality of life* should be provided by HCPs to patients, ensuring that patients are fully aware of and educated on the relationship between FC, a broader and more subtle definition of quality of life and goals for clinical care
   - The impact of a decline in FC on patients’ quality of life* should be assessed and taken into account during patient review and treatment planning

CALL TO ACTION FOR CARER

Caring for a patient or loved one with PAH is a huge undertaking and one that can affect physical, practical, social, emotional and financial aspects of life. It is, therefore, imperative for carers to fully understand these impacts and gain support and guidance to ensure their and the patient’s quality of life are not compromised as a result of their carer role.

1. Carers should be able to request access to HCPs and patient organisations who have a strong understanding of the carer burden and the important role the carer plays for the patient
   - Caring for a patient with PAH has a major impact on the carer and this should be recognised by all members of the team providing the clinical care
   - Carers’ needs are often less visible than the patients’ and are, therefore, frequently overlooked
   - The financial burden of PAH necessitates more external support for carers, who are often responsible for the administrative consequences of disease

2. Information needs of the carer are often different to the patient and must be urgently addressed
   - Carers require tailored information to help them care for someone with PAH (including advice on social, economic and financial aspects) and information for carers should be addressed to carer needs specifically
   - Carers are encouraged to be proactive in seeking more support from patient organisations, HCPs and PAH support networks

* Quality of Life not based on the standardised instruments used in Patient Reported Outcomes