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.

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* A selection of questions asked during the survey can be found throughout this report

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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, internists, 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.

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 (35% had felt this in the last month); feeling misunderstood; little pleasure in activities; anger and low self-esteem.

Nearly 25% of carers 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 should be addressed, including the provision of information on social, financial and emotional impact, as well as on PAH disease and its treatment. The findings also identify these areas for further research to improve the understanding and overall management of PAH.

*Participants were not asked for net or gross income so results assume after tax

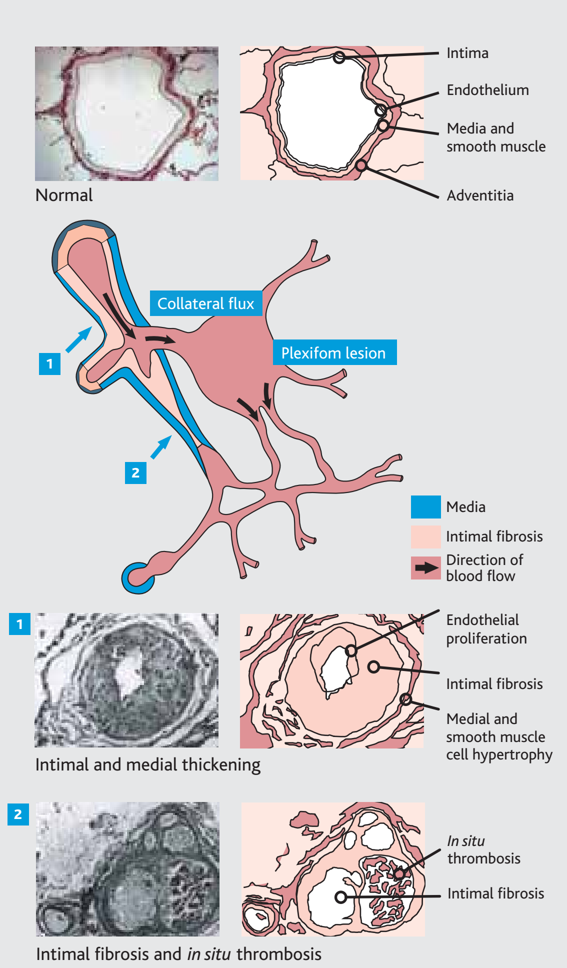
WHAT IS PAH?

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-1980s there were limited treatment options for patients and PAH was associated with poor prognosis.

Illustration 1. **PAH: histopathological features**



www.PAH-info.com

HOW COMMON IS PAH?

Although PAH is a rare disease, with an overall estimated prevalence of 15-50 cases per million[3], 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 CAUSES PAH?

One of the more common forms of PAH is idiopathic PAH (IPAH), which is a sporadic disease in which there is neither a family history of PAH nor an identified risk factor.[4] Heritable PAH (HPAH) accounts for at least 6% of cases of PAH[5] and mutations in the bone morphogenetic protein receptor 2 (BMPR2) have been identified in the majority of cases.[6] PAH can also be caused by the use of certain anorexigenic agents, such as fenfluramine.[2,7] However, the incidence of drug-induced PAH related to fenfluramine is decreasing since this agent is no longer available.

PAH can also be associated with a number of other conditions (associated PAH, APAH), which together account for most other cases. These conditions include: connective tissue disease; HIV infection; portal hypertension; congenital heart disease; schistosomiasis and sickle cell disease.

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.[2,7,8] 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.

> *It totally changes your life* - you can't ignore it because it will worsen and beat you until you're unable even to turn and move a pillow. *Your body is weak*, you have to pace yourself so as not to use precious energy. **Patient**

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).

Table 1. **WHO functional classification of pulmonary hypertension** [2]

FUNCTIONAL CLASS	SYMPTOMATIC PROFILE
I	Patients with pulmonary hypertension but without resulting limitation of physical activity. Ordinary physical activity does not cause dyspnoea or fatigue, chest pain, or near syncope
II	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
III	Patients with pulmonary hypertension resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnoea or fatigue, chest pain, or near syncope
IV	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/or fatigue may even be present at rest. Discomfort is increased by any physical activity

PAH currently tends to be evaluated based on a physical definition of clinical symptoms including clinical evaluation, exercise tests, biochemical markers and echocardiographic and haemodynamic assessments.[2] For example, exercise capacity is measured using the six-minute walk test (the distance that a patient can walk in six minutes). However, PAH has a much broader impact than suggested by the current definition of clinical symptoms but there has been relatively limited previous research into this.

IMPACT OF PAH ON CARERS

The limitations imposed by PAH mean that patients often need help from a carer, with partners and family members taking on this role. Becoming a carer can have a major effect on a person's life, often reducing their opportunities for working and taking part in other activities. However, the needs of carers of PAH patients are often overlooked and they are not even mentioned in the European or US guidelines on pulmonary hypertension [2, 9], in contrast to guidelines concerning other long-term serious conditions such as cardiovascular disease and Alzheimer's disease where the role of carers is increasingly being recognised and they are offered help and support.[10, 11]

> 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 150kmph* because all she wanted was to get home. **Carer**

> I would say *my own emotional journey is more or less normal* but does go up and down slightly. I am content now because I have a great trust in the transplant. **Carer**

METHODOLOGY

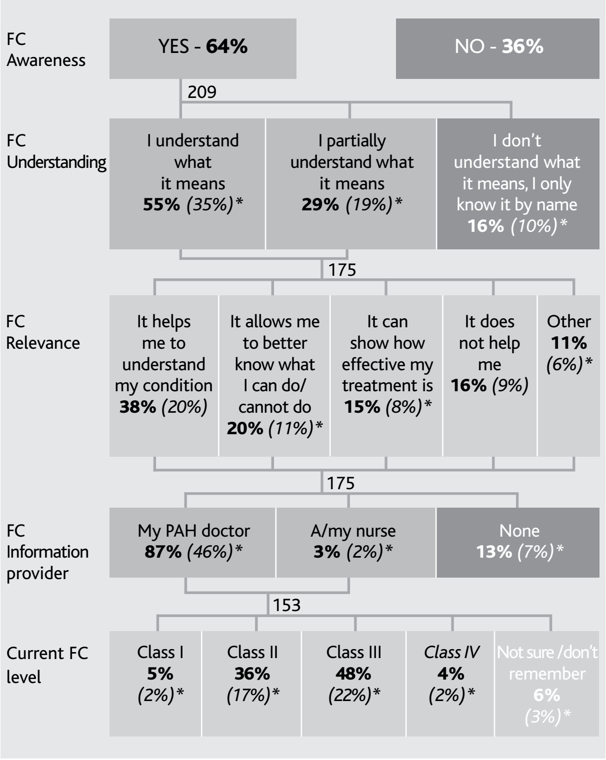
This research was set out to explore four main areas: physical and practical impact of PAH; emotional impact; social impact; and information needs and provision; using market research methodologies (Table 2).

Table 2. **Schematic representation of questionnaire**
Total: 59 questions, which were mostly with multiple choice possible responses

Respondents' details including background information on PAH:	GENERAL INFORMATION ↓			
Impact of PAH on day-to-day life:	**Physical** (including work & intimacy)	**Practical** (including work & income)	**Social** (including friends & family)	**Emotional** (including underlying feelings & support)
Other PAH related topics:	**Provision of Information** (including sources, topics & timing post diagnosis)			

* For emotional impact, questions focus on how frequently the respondent experienced a range of listed emotions over the last month ranging from "never" to "very often (almost every day)"

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



FC Awareness	YES - **64%**	NO - **36%**			
	209				
FC Understanding	I understand what it means **55%** *(35%)**	I partially understand what it means **29%** *(19%)**	I don't understand what it means, I only know it by name **16%** *(10%)**		
	175				
FC Relevance	It helps me to understand my condition **38%** *(20%)*	It allows me to better know what I can do/ I cannot do **20%** *(11%)**	It can show how effective my treatment is **15%** *(8%)**	It does not help me **16%** *(9%)*	Other **11%** *(6%)**
	175				
FC Information provider	My PAH doctor **87%** *(46%)**	A/my nurse **3%** *(2%)**	None **13%** *(7%)**		
	153				
Current FC level	Class I **5%** *(2%)**	Class II **36%** *(17%)**	Class III **48%** *(22%)**	*Class IV* **4%** *(2%)**	Not sure /don't remember **6%** *(3%)**

* Percentages in brackets are percentages of the total sample

It was carried out in two phases:

- Phase I was a qualitative survey based on one-to-one interviews between 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 *
  *All patient and carer quotes listed were collected during Phase I research
- 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]) 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 that each country 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, internists, 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.

1.0 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 describes the impact of your PAH on your daily life?

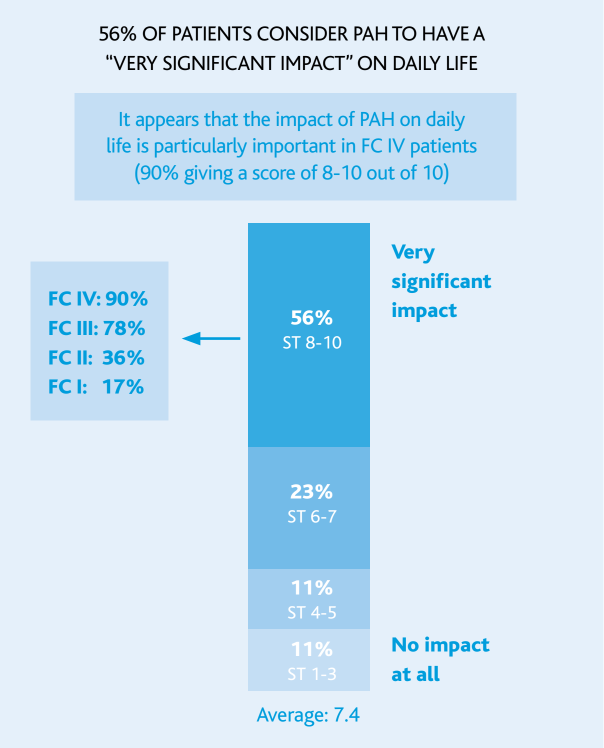
Patients were frequently unable to perform everyday or 'normal' domestic tasks, however small, such as household chores and shopping for food; 56% of patients said that PAH had a 'very significant' impact on their daily life, while a further 23% said it had a 'moderate impact'.

> She manages to cook at times, but *right now can't really manage that at all*. **Carer**

> 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 10kph*. **Patient**

Chart 1. **How patients assess the impact of PAH on their daily lives**



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:

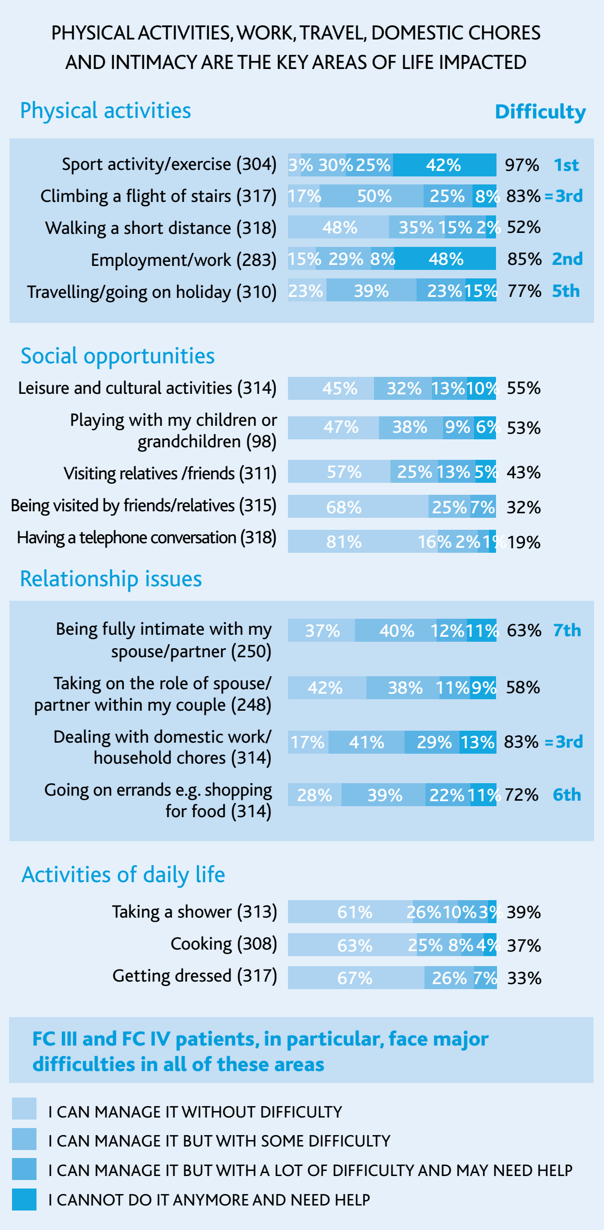
- Physical activities – overall, 97% of participants taking part in the survey found their ability to take part in sports and exercise was affected; 83% had difficulty climbing stairs; 52% of patients had more difficulty walking a short distance compared to before their diagnosis (Chart 2)
- Employment/work – overall, 85% of patients found their employment/work was affected by their condition
- Travel and social opportunities – overall, 77% of PAH patients reported that their ability to travel or go on holiday was impacted; 55% said leisure and cultural activities were

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**



The proportion of patients reporting difficulty with all of these activities increased with decline in FC.

1.1 IMPACT ON EMPLOYMENT/WORK AND INCOME

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 overall household income, to 67% of those with FC II, 79% of those with FC III and nearly all (95%) of patients with FC IV.

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*Participants were not asked for net or gross income so results assume after tax.

1.2 IMPACT ON SEX LIFE AND ABILITY TO HAVE A FAMILY

In the qualitative interviews, when asked, almost all patients reported that their sex lives were severely affected, mainly because of low self-esteem and being unable to physically exert themselves. In the survey, 44% of patients reported experiencing loss of libido/interest in sex often or very often in the previous month (time period asked about in the survey).

Intimate relationships were adversely affected as FC declined, with 66% of patients with FC IV experiencing a loss of libido 'often to very often', compared to 57% of FC III patients, 36% of those with FC II and none of the patients with FC I.

> I don't feel great about my body with a big tube sticking out of it. It's not what you would call sexy. **Patient**

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 identify was reduced by not being able to perform maternal tasks with their children and housework.

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**

> Nobody understands my suffering and they don't want to listen. Everyone, doctors especially, even my daughter. **Patient**

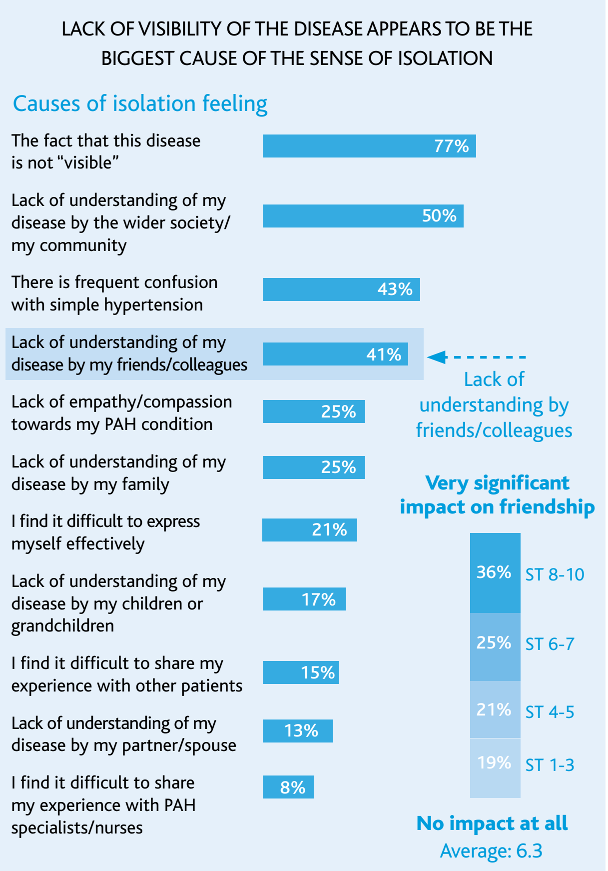
> My friend said 'I wish I had your kind of hypertension' and I was so upset by that. She thinks I am at home and just sitting around all day. **Patient**

Many patients (n=178; 40% of the sample) reported experiencing a feeling of isolation in the last month, which was the time period they were asked about in the survey (Chart 3). Of these, 77% considered this was caused by PAH not being 'visible,' which was particularly significant in younger patients (100% of patients under 30). A high number of patients felt that a lack of understanding among family and friends contributed to their isolation. One in four patients (25%) found a lack of understanding of their disease by their family, 17% by their children or grandchildren and 13% by their partner/spouse. Under half (41%) of patients experienced a lack of understanding among their friends and colleagues, and one-third of these patients (36%) said this had a very significant impact on friendships.

Half (50%) felt their isolation was because of lack of understanding by the wider society or their community.

> *Can be isolating,* sometimes I have *no desire to socialize* - sometimes I don't want to go out and circulate and talk to people. **Patient**

Chart 3. **Causes for feelings of isolation as a result of PAH/impact on friendships**



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 women, young patients and 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 FC, 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 changed 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 patients' 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 felt 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.

3.0 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 uselessness, 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 this 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, hopeless, afraid to leave home, guilt, no desire to socialise, feeling embarrassed, fearful at night and fearful during the day. Just under half (41%) of patients had experienced three or more of these feelings often or very often in the last month.

Many patients showed signs associated with depression, including isolation, feeling low in the mornings, and feeling hopeless, but most did not recognise or label themselves as being 'depressed' per se. Signs and symptoms that are commonly associated with depression were not only reported by many patients in FC III and IV, as would probably be expected, but also, to a significant extent, by patients in FC II. This emphasises the importance of not underestimating these feelings among patients early in the disease progression.

> I was really low when I was diagnosed - I thought I was going to die. I used to wake up and have panic attacks being just really frightened, feeling really vulnerable, I was feeling crap. I would wake up with night terrors and I couldn't stop screaming. **Patient**

> What *made me feel depressed was* when they told me I had to *stay in hospital*, I had never been in hospital before. **Patient**

> It's not a good feeling - it's bad to be dependent on others. It is bad to ask other people to help you. You feel bad. You feel inferior - like a second class citizen. **Patient**

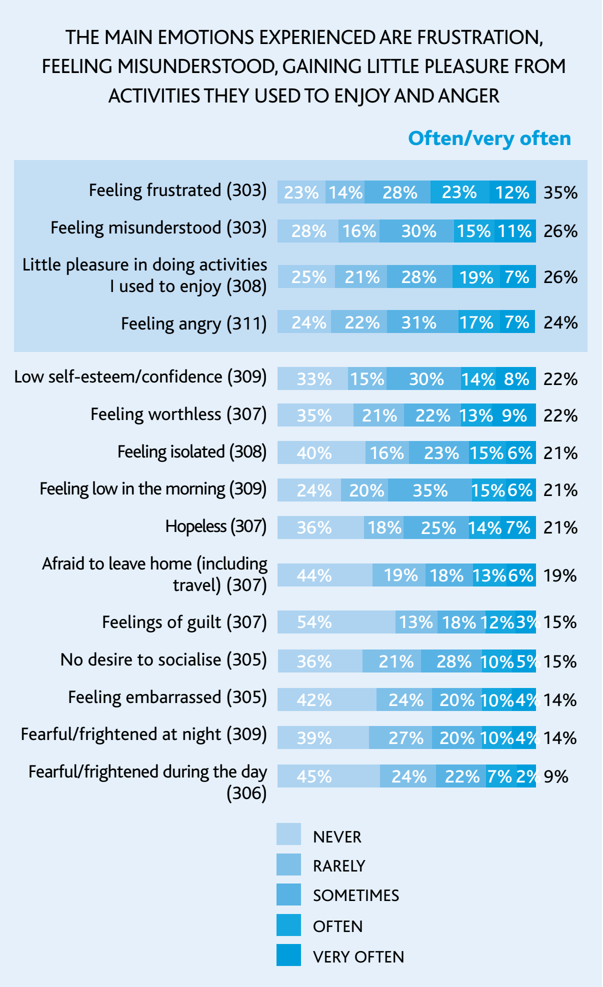
Who is providing you with emotional support about living with PAH?

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?

Patient organisations are among the top three places where patients found discussions about feelings are easier and 30% of patients said they provided them with support. However, only 45% of respondents claimed they had been pro-actively referred to patient organisations by specialists (despite European guideline recommendations[2]), indicating a need for greater education amongst PAH specialists about the role of patient organisations and the importance of working with them collaboratively.

Chart 4. **Frequency of emotions in the last month**



EMOTIONAL IMPACT OF PAH ON PATIENTS

Although the physiological needs such as breathing are at the front of the mind for patients, they are undoubtedly experiencing a wide range of feelings 'beneath the surface'. When probed, their frustration, stress, hopelessness, low mood and lack of self-worth are given voice. This points to a need for patients to be provided with and/or directed to good and appropriate emotional support, for example, towards the patient organisations.

4.0 KEY RESULTS: PATIENTS' INFORMATION NEEDS

In the qualitative interviews, patients reported that they had been given very little information on the emotional impact of PAH by HCPs 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. Patient organisations were the most important source of information for patients, being seen as trustworthy and credible, but results suggest they could provide more information on social and financial support.

> The patient organisation keeps you up to date with things and you're not alone. **Patient**

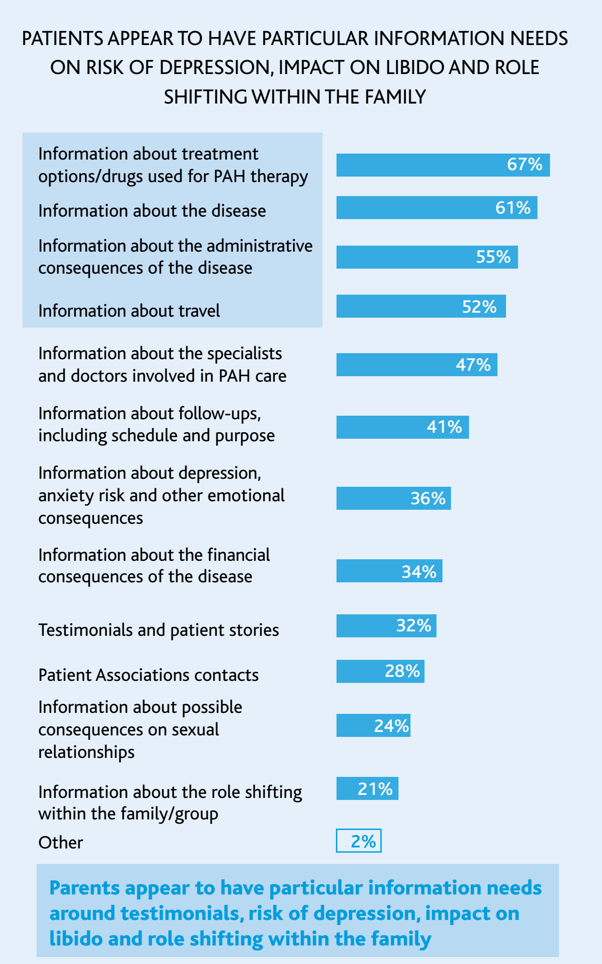
Where are you likely to go and look for information about PAH?

In the quantitative survey of patients, patient organisations emerged as one of the most important sources of information for patients*, with 66% of patients looking for information about PAH from them, and 30% gaining emotional support from these organisations.

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

Patients' top information needs were: treatment (67%), the disease (61%), the administrative consequences (55%), travel (52%) and doctors involved in PAH care (47%) (Chart 5). Over one-third wanted information about depression and other emotional consequences, and a similar proportion wanted patients' stories and patient association contacts.

Chart 5. **PAH information required by patients**



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* The majority of patients and carers were recruited via patient organisations and were current members of patient organisations members (a mixture of active and non-active)

Please indicate the most suitable time for you to be given the following information?

Most patients wanted key information at diagnosis – particularly about the disease and its treatment. Information on consequences such as administrative and financial aspects can be provided at a later date (i.e. 1-3 months after diagnosis and as far as 1 year post diagnosis and beyond) (Table 4).

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 (ESPECIALLY ON DISEASE AND TREATMENTS), WHILE OTHER TOPICS BECOME MORE IMPORTANT SUBSEQUENTLY

INFORMATION ABOUT…	% PATIENTS WHO WISH TO RECEIVE	AT DIAGNOSIS	1 MONTH AFTER	3 MONTHS AFTER	6 MONTHS AFTER	1 YEAR + AFTER
Treatment options/drugs	67%	80% 2nd	12%	4%	1%	3%
The disease	61%	83% 1st	9%	4%	1%	3%
The administrative consequences of the disease	55%	40%	28%	16%	10%	6%
Travel	52%	24%	21%	25%	16%	14%
Specialists and doctors involved on PAH care	47%	77% 3rd	12%	5%	3%	3%
Follow-ups including schedule and purpose	41%	58% 4th	25%	11%	3%	3%
Depression anxiety risk and other emotional consequences	36%	47%	26%	14%	7%	6%
Financial consequences	34%	38%	31%	17%	7%	7%
Testimonials and patient stories	32%	26%	25%	24%	11%	14%
Patient Association contacts	28%	50% 5th	17%	16%	6%	11%
Sexual relationships	24%	40%	25%	21%	7%	7%
Role shifting in family/couple	21%	39%	29%	19%	6%	7%

> On the site *I read about patients who had managed to lead almost normal lives* so found this very helpful. **Patient**

> One of the first things it (internet) said was I could live for 2.5 years with PAH - not pleased to read this straight after diagnosis! **Patient**

PAH INFORMATION NEEDS: THE IMPACT ON PATIENTS

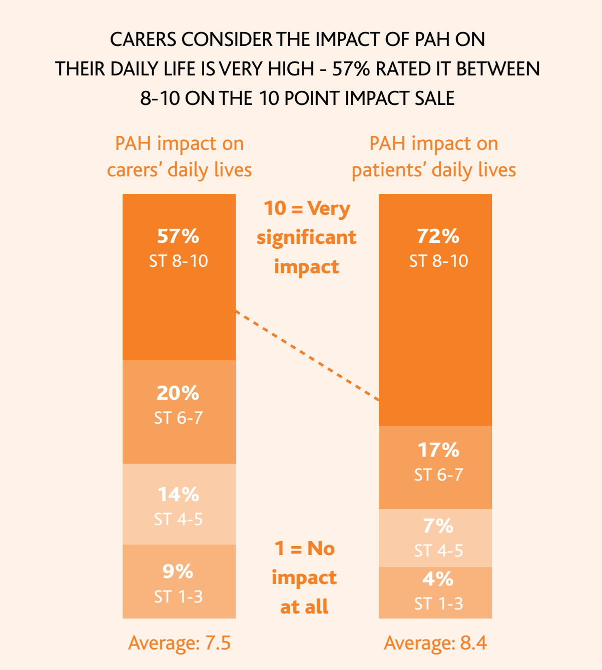
Patients report that little information is given at diagnosis and they are left with unanswered questions. Many look to the patient support organisations for their information and emotional needs to be met but many patients are not directed to this important source of support by their HCPs. Patients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

1.0 KEY RESULTS: PHYSICAL AND PRACTICAL IMPACT ON CARERS

The qualitative interviews showed that carers take on a 'fighter' and 'organiser' role, carrying out physical tasks and chores to compensate for patients' inabilities 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)**



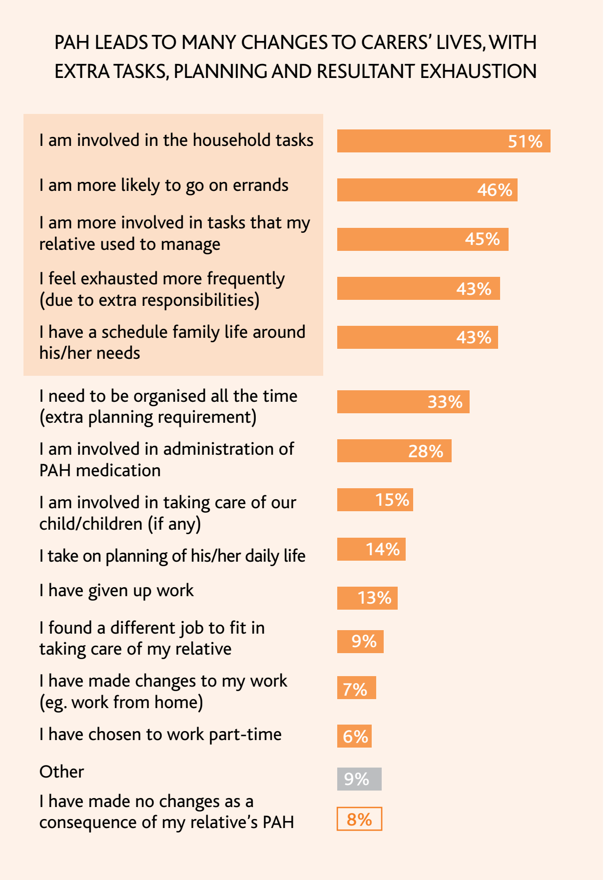
Carers rated the overall impact of their relatives' PAH on their daily lives as very high: 57% rated it between 8-10 on a 10-point impact scale (Chart 6). A higher proportion of carers of patients of FC III and IV reported a very high impact on their lives (53% and 88%, respectively) than those caring 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% were more likely to 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: 43% 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.

> I'm very much involved and I've taken on a lot of household chores. I clean windows, vacuum and cook because the person I care for is not able to do this any longer. **Carer**

Chart 7. **Changes to carers' lives as a result of PAH**



Has there been an overall decrease in YOUR sexual relations?

Nearly three-quarters (72%) of carers who are partners of PAH patients reported a decrease in sexual relations, mainly due to the patient's lack of interest since having PAH and their own fear of making the patient more ill. The impact on sexual relations increased with a decline in FC, with a significant effect at FC II (57%), increasing to 74% at FC III and affecting virtually all carers (93%) caring for PAH patients at FC IV.

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.

PHYSICAL AND PRACTICAL IMPACT OF PAH 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.

2.0 KEY RESULTS: SOCIAL IMPACT OF PAH ON CARERS

During interviews, carers revealed that their world narrows because they spend much of their time caring for someone with PAH and they become restricted socially, although do not appear to suffer the same loss of social identity as the patient. They considered there was a lack of public knowledge and awareness of PAH, which affects how people view patients with the condition.

In a normal month, how much time do YOU spend, on average, doing the following?

The survey results showed that half (51%) of carers' time, on average, is spent caring for someone with PAH in a normal month. This represented a sharp 20% increase compared to before the person they care for was diagnosed with PAH.

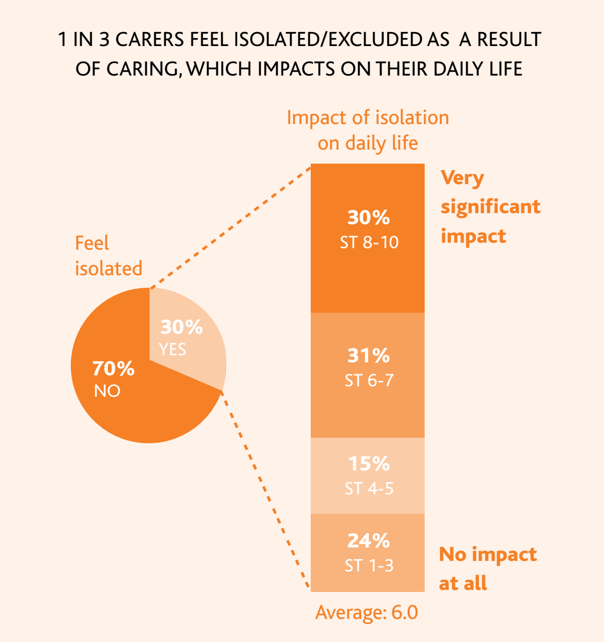
The time spent caring for someone with PAH increased as FC declined (FC II 49%; FC III 62%; FC IV 70%).

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**



> 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

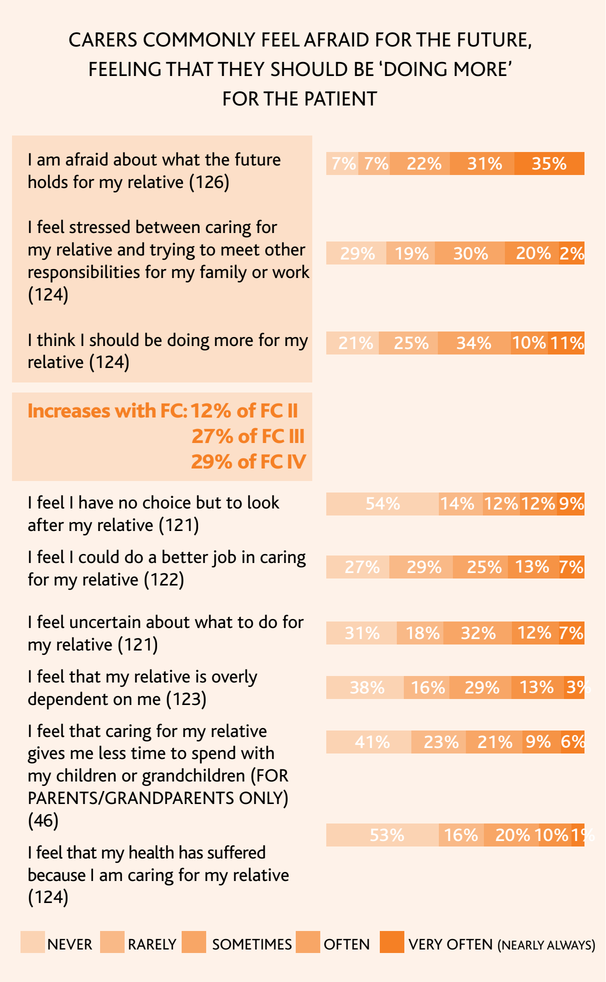
The research highlighted that caring for someone with PAH can have a significant impact on the carers own social life. Carers are restricted socially as they spend a high proportion of their time caring, which can result in feelings of isolation. The time spent caring increases as the functional class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

3.0 KEY RESULTS: EMOTIONAL IMPACT ON CARERS

The in-depth interviews suggested that carers tend to prioritise the patient's emotions above their own. But 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**



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.

> Living with someone with PAH is like not knowing what to expect, I don't know how it will develop. This is the first thing I think about, as a parent, not seeing him grow up and develop. **Carer**

> 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. **Carer**

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 carers (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.

> *Depressed* - this might be an exaggeration - but she gets sad. *She gets sad* around the time of her check ups, having to spend two hours in the car, and thinking about hospital...
>
> Every check up *she is terrified it has got worse...* now we have got to the transplant, which is the last card to play...
>
> *She lives in the fear everyday* - that she will get worse again and the treatment will stop being effective. **Carer**

EMOTIONAL IMPACT ON CARERS

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.

4.0 KEY RESULTS: CARERS' INFORMATION NEEDS

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.

> I use the internet continually, but I have to filter internet information a lot. I have to consider whether the information I will pass on to my wife is true, and relevent, so scientifically based information. **Carer**

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%).

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.

DISCUSSION AND CONCLUSIONS

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 experience when living with PAH, with just over half of patients and carers taking part in the survey saying that the condition has had a ‘very significant’ impact on their daily life.

Detailed questionnaires to 326 PAH patients and 129 carers from five European countries - France, Germany, Italy, Spain, and the UK – revealed the serious limitation that PAH imposes on ability to perform everyday tasks, however small, such as household chores, shopping for food or taking a shower. The key areas of life impacted by PAH emerged as physical activities, with over half of patients having difficulty in walking a short distance, employment and work, travel and social opportunities and relationship issues. Carers had to take on more household tasks and activities that their relative used to manage. Unsurprisingly, a decline in FC was associated with a greater impact on daily life for the patients themselves and for carers.

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 half saying they were unable to work anymore or needed help to do so because of their condition. Capacity to work was also affected, with nearly one in three of those 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, leaving most families below the average level of income.

Relationship issues emerged as a previously under-appreciated area of concern for PAH patients and their carers. Almost all patients taking part in the in-depth qualitative interviews reported that their sex lives were severely affected by their condition, mainly because of low self-esteem and being unable to exert themselves physically. Loss of libido was also confirmed in the quantitative survey. Carers who are partners of PAH patients were also obviously affected by this, with three-quarters reporting a decrease in sexual relations, mainly due to the patient’s lack of interest since being diagnosed with the condition and their own fear of physical activity making the patient more ill. In turn, this reduction in sexual relations impacted on their ability to consider having children.

The interviews and surveys revealed how much the world narrowed for both PAH patients and carers because of being much more confined to the home and less able to travel and socialise. This was manifested as the loss of many of the social roles that they once held, including those as a colleague in the workplace, and as a friend. Many patients and carers reported feeling isolated, often caused by their PAH not being ‘visible.’ A surprisingly high number of patients felt that lack of understanding of their condition among family and friends contributed to their isolation. Improved provision of information on PAH tailored for patients, carers and family and friends specifically, would be useful in helping to bridge this understanding gap.

The challenges imposed by living with PAH exacted an emotional toll on patients and carers. 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.

LOOKING TO THE FUTURE

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 psychological 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 about 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, emotional and social – so they can enjoy the best outcomes and quality of life that are achievable.

CALL TO ACTION FOR HEALTHCARE PROFESSIONALS

Comprehensive care is essential to improve the practical, physical, emotional and social well-being of PAH patients and carers. HCPs should extend assessment and care beyond the clinically defined symptoms of the disease.

1. HCPs to assess patients beyond the clinically defined impact of PAH and include the practical, physical, emotional and social impact

- It is important to emphasise that the disease burden of PAH (practical, physical, emotional, and social) increases significantly with a decline in FC
- Treatment guidelines need updating to provide more comprehensive care for patients and carers beyond the clinically defined symptoms of the disease, and to include practical, emotional and social aspects
- The impact of a decline in FC on patients’ quality of life* should be assessed and taken into account when planning treatment
- HCPs need to better understand the wider impact of PAH, in particular the financial impact it has on patients and carers

2. Optimal PAH care requires a multidisciplinary approach for patients and carers

- Patients and carers should be offered comprehensive, coordinated care from a multidisciplinary team of HCPs and other specialists, including physicians, nurses, social workers, psychologists and patient organisations
- Clinical care should be adapted to the individual patient, including consideration of their physical and emotional needs
- The needs of the carer are currently underestimated and often overlooked; their needs should be addressed as part of the patient’s multidisciplinary package of care

3. HCPs need to provide patients’ and carers’ information needs in a timely way, which often needs to be repeatedly provided to meet their needs

- Patients and carers require different information at different stages after initial diagnosis. Information should be provided in stages when the patient and carer need it, which may mean giving information on PAH and treatment at diagnosis and on practical, emotional, social and financial aspects at a later stage

4. Relationships should be strengthened between HCPs and patient groups

- Patients under specialist care and their carers should be directed to patient groups to gain further support and access to the PAH community
- HCPs and patient groups need to develop stronger links to ensure development and timely provision of appropriate PAH information on a range of topics beyond only the physical impact, including patient/carer information resources on financial, emotional, social and physical impacts of PAH, PAH/PAH symptom awareness campaigns and patient referral to appropriate patient support groups at specific times in disease progression

CALL TO ACTION FOR PATIENT ORGANISATIONS

The role of PAH patient organisations has been identified as integral in the effective management of PAH. Patient organisations should ensure that PAH assessment and management extends beyond the clinical symptoms of the disease; providing a high level of support including information provision and emotional support, which both supplements and improves the standard of PAH care provided

1. Strengthen relationships between patient organisations and HCPs

- HCPs and patient organisations need to explore ways to improve dialogue and collaboration. Patients and carers should be referred to patient organisations as part of routine provision of good care
  - Patient organisations need to support patients and carers to more confidently communicate the burden of PAH – including the wider aspects such as emotional, social and financial impact - and ask for the help they need

2. Increased recognition of the carers within membership

- Patient organisations need to ensure that adequate provision is in place to support the carers of their patient members

3. Increase awareness of non-physical impact of PAH

- Patient groups should campaign for greater support for patients, including the need for financial help, emotional support and support when returning to work
- They should provide information sessions and educational events to help to improve members understanding of PAH and its impact not only on the patient, but also on family and friends

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

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* (broadly defined) should always 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 theirs and the patients' 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 pro-active in seeking more support from patient organisations, HCPs and PAH support networks

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

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