PAH PATIENT CHARTER

Pulmonary arterial hypertension (PAH) is classed as a type of pulmonary hypertension (PH). PH is a wider term used to describe the presence of high blood pressure in the lungs due to any cause. PAH is caused by the walls of the arteries in the lungs becoming tighter and narrower.[1]

The PAH Patient Charter is based on the most current (2015) European Society of Cardiology/European Respiratory Society (ESC/ERS) guidelines[2], which are used internationally, and was developed from the outputs of a meeting held in Berlin, Germany, in March 2019. The meeting participants included patients, patient advocacy group representatives and healthcare professionals from the PAH global community; the content of the Charter was informed entirely by the discussions, reflecting their own views and perspectives on the best holistic care package for people with PAH.

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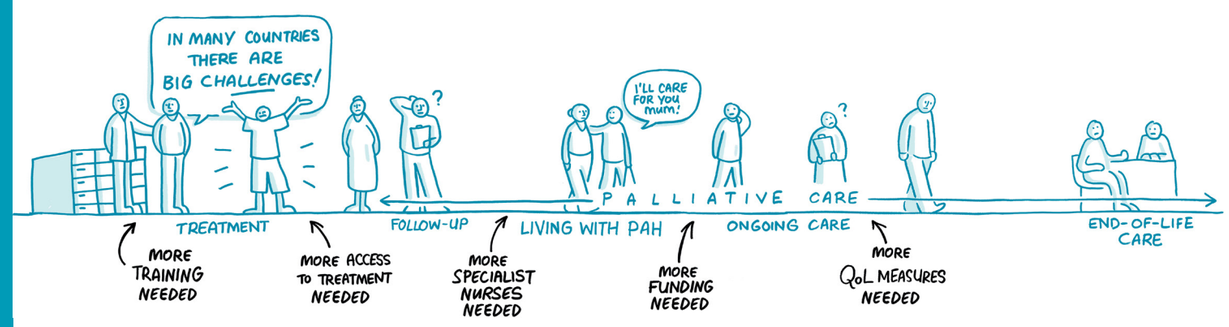
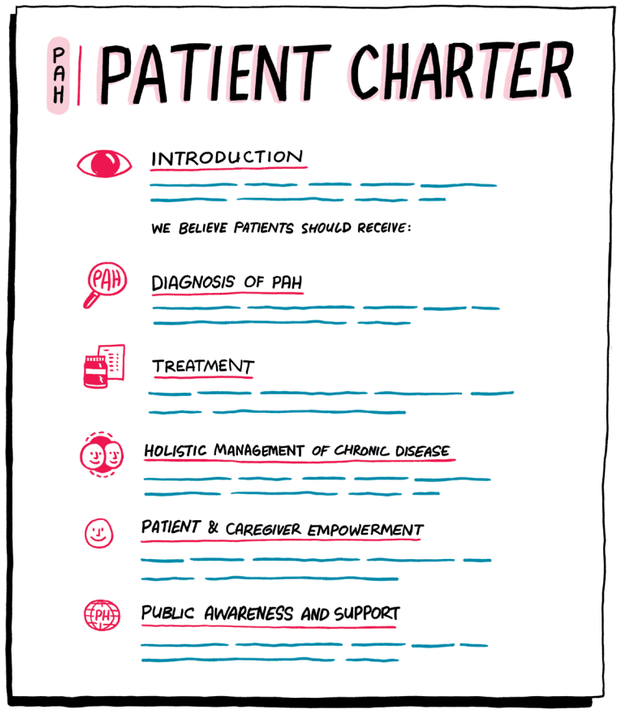
WHAT YOU SHOULD EXPECT FROM YOUR CARE



The cartoons throughout this Charter were drawn during the Steering Committee meeting at which they discussed its content. Each cartoon reflects parts of those conversations, but may not correspond specifically with the content of the page it is on.

HOW TO USE THE PAH PATIENT CHARTER

The PAH Patient Charter outlines the standards of holistic care that we believe everyone with PAH should receive.



It is based on ESC/ERS international guidelines[2] on the diagnosis and treatment of PAH and has been developed by an international group of patients, patient advocacy group representatives and healthcare professionals.

We hope that the information in the Charter will help you, your healthcare professionals and caregivers to:

- Communicate effectively with each other
- Make decisions together
- Agree on a care plan that is appropriate for you

If you think that you are not receiving the kind of care described in the Charter, there are patient advocacy groups, otherwise referred to as patient associations or patient organizations, in many countries that may be able to help you. In fact, the Pulmonary Hypertension Association (US) provides a directory of approximately 90 PH associations worldwide.[3] Patient advocacy groups can provide you with the opportunity to speak to other people living with PAH, who can be difficult to find and connect with on your own. As a newly diagnosed patient, for example, it may be very helpful to speak to someone who has lived with PAH for a long time, for tips and motivation. Patient advocacy groups can provide you with guidance on the type of care and social benefits available in your country. Patient advocates can also provide you with an understanding of how to navigate the care system in your country based on their own experience.

YOU SHOULD EXPECT TO RECEIVE A TIMELY AND ACCURATE DIAGNOSIS

PAH is a rare disease with relatively low awareness amongst the public and healthcare professionals.

As its symptoms can be similar to those of more common conditions like asthma and chronic obstructive pulmonary disease (COPD), it can take an average of two years from the onset of symptoms to diagnosis in countries with developed healthcare systems.[4] This means that people with PAH are often seen by several doctors and nurses, including a cardiologist (heart specialist) and a pulmonologist (lung specialist), before they receive a diagnosis.

Whilst it is not always realistic to expect a rapid diagnosis following onset of symptoms, in countries with developed healthcare systems, a feasible 'timely' diagnosis should be significantly less than the current two-year average.

If you have been told by your doctor that they suspect that you may have PAH in view of your symptoms you should expect to be offered tests in line with applicable diagnostic guidelines to confirm whether you have the condition. Four important tests used in the process to diagnose PAH are:[5,6]

- An echocardiogram: an ultrasound exam that shows how well your heart and nearby blood vessels are working
- A ventilation/perfusion (V/Q) scan: a scan that measures the distribution of air and blood in your lungs. This test can rule out forms of PH due to clots or scarring from clots. This type of PH is called chronic thromboembolic pulmonary hypertension (CTEPH)
- Computerized tomography pulmonary angiography: a scan that provides an image of the pulmonary arteries, allowing your doctor to identify any blockages in the vessel. Some experts use this as an alternative to a V/Q scan, although V/Q scans remain the preferred test for screening
- A right heart catheterization: a procedure to see how well your heart is pumping, and to measure the blood pressure in your heart and the blood vessels leading to your lungs. This is considered the gold standard in diagnosing PAH and is always required to confirm a diagnosis of PAH

You should expect your healthcare professionals to explain what the diagnostic tests you are being offered involve. Once you have had the tests, you should expect your healthcare professionals to explain what the results mean and what they think the next steps should be.

If the tests indicate that you have PAH, you should expect your healthcare professionals to explain the diagnosis to you and give you the opportunity to ask questions. You should expect to be able to have a family member or a friend with you if you want them to be there.

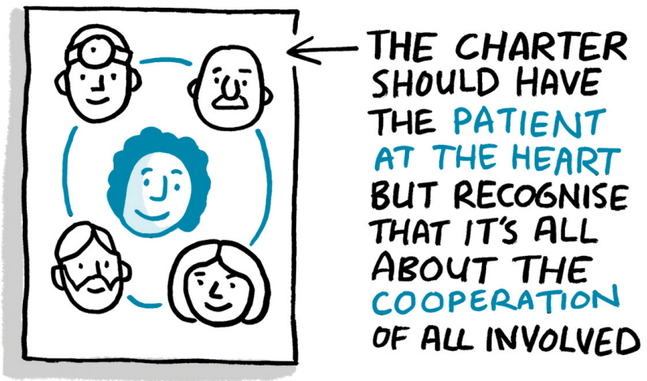
People who assist PAH patients with different aspects of their care, such as their daily treatment routine, go by different names – carers, caregivers, and care partners. There may also be resources available to them from your healthcare system, patient advocacy group, caregiver organization, or government agencies.

If the tests were not conducted in a specialist PAH treatment center, and such centers exist in your country, you may be referred to one once PAH is suspected. Specialist PAH centers are not available everywhere, but your patient advocacy group, healthcare system staff or other authority may be able to inform you of their existence. As it is a rare condition, only select healthcare professionals will have in-depth knowledge about PAH and its treatment. As such, you should expect a PAH specialist to oversee your care.

If there are several healthcare professionals involved in your care, you should expect them to communicate effectively with you and each other.

YOU SHOULD EXPECT YOUR HEALTHCARE PROFESSIONALS TO LISTEN TO YOUR PREFERENCES AND INVOLVE YOU IN DECISIONS ABOUT YOUR CARE AND TREATMENT

This means that you should expect your healthcare professionals to ask you about what is important to you and what you want your care and treatment to achieve.



They should talk to you about the different kinds of care and treatment that are available and appropriate based on expert consensus guidelines, so that you can make an informed decision together about the care plan that is the most appropriate for you. This should include conversations about multidisciplinary symptom management and holistic or palliative care for you or your loved ones.

You should expect your healthcare professionals to ask you about how you are coping with the care plan to which you have agreed. It is important to remember that only you know how you are feeling. Expect referrals to psychosocial support professionals if those are available in your healthcare system.

If your healthcare professionals suggest any changes to your care plan, they should explain what changes they are suggesting, the reason for the proposed changes and what impact they expect the changes to have. They should also communicate changes to other members of the care team such as social workers, pharmacists, other physicians and care partners.

YOUR HEALTH SHOULD BE REGULARLY ASSESSED AND YOU SHOULD BE INFORMED ABOUT THE FULL RANGE OF AVAILABLE, EVIDENCE-BASED TREATMENT OPTIONS

You should expect your healthcare professionals to conduct a risk assessment to help them set a holistic care plan that is most appropriate for you. A risk assessment will involve some tests and labs to assess your functionality and to measure how well your heart and lungs are working. As PAH is a progressive disease, you should expect your healthcare team to conduct a risk assessment at each visit, about every three to six months for most patients, in order to evaluate your current care plan.[2]

After your risk assessment you should be informed by your healthcare professionals of the results of your assessment and about the full range of available, evidence-based treatment options.

There are a large number of licensed medicines that are recommended for the treatment of PAH, though not all of these medicines may be available in your country.

You should expect your healthcare professionals to explain why they are recommending each medicine they prescribe and the benefits and risks that they might have. For example, they should explain the common side-effects and how the medicine might make you feel. If you have had a risk assessment before, you should expect your healthcare professionals to explain whether your health has changed since your last assessment.

TREATMENT SUMMARY

There are four types of PAH-specific drug treatments, known as:[2]

- Endothelin receptor antagonists (ERAs)
- Prostacyclin pathway agents
- Phosphodiesterase 5 (PDE-5) inhibitors
- Soluble guanylate cyclase stimulators (sGCS)

These medicines relax and widen the blood vessels in the lungs as well as reduce and prevent the overgrowth of cells in the walls of the vessels, making it easier for the heart to pump blood through the lungs.[7]

For a very small number of patients that demonstrate a specific and positive response to part of the right heart catheter diagnostic procedure, calcium channel blockers (CCBs) may be recommended as initial therapy, in the short-term.[2,8] These are a group of medicines – not specific to PAH – that also work by relaxing and widening blood vessels.[8]

There are several other treatments that may be considered which can help prevent specific consequences or relieve symptoms of PAH, including:[2]

- Blood thinners to prevent clots (eg, warfarin)
- Diuretics (also called water tablets) to reduce swelling
- Supplementary oxygen to ensure the body has enough oxygen

Availability of PAH treatments varies by country and locality.

YOU SHOULD HAVE ACCESS TO HOLISTIC CARE AND SOCIAL SUPPORT

You should expect your healthcare professionals to direct you towards information and support services that will help you and your family live well with PAH.

For example, this might include information and support for your mental health and emotional wellbeing, as well as advice on a wide range of topics such as travel, nutrition, family planning, physical activity, career choices, financial planning and access to psychological support.

If others are involved in your care, then you should expect them to be offered information and support to help them in their role as caregivers.

You should expect your healthcare professionals to provide you with information about your local PAH patient advocacy group, or another related patient advocacy group. A patient advocacy group may be able to put you in touch with other people living with PAH as well as provide you with information and support. They should also be able to advise you on what social benefits you may be able to receive from your country or state, including whether you have the right to disability status.

CALLS TO ACTION

We hope that this PAH Patient Charter will help you and your healthcare professionals communicate effectively with each other, make decisions together and agree on a care plan that is appropriate for you.

The patient advocacy groups endorsing the Charter are also working to ensure that everyone with PAH receives the same standard of holistic care outlined in the Charter. Together we are calling on healthcare systems to:

DIAGNOSE PAH EARLIER

- Provide healthcare professionals with opportunities to learn about PAH, from medical school to ongoing medical education, to help them recognize the signs and symptoms of the condition
- Educate and encourage healthcare professionals to consider assessing the right heart in cases of unexplained breathlessness
- Introduce clear protocols for diagnosing PAH, as outlined in ESC/ERS international guidelines[2]
- Ensure that specialist PAH centers exist, and that they have the resources and the capacity they need to diagnose PAH early
- Collect and report data on how long it takes for people with PAH to receive a diagnosis after their first visit to a healthcare professional

SUPPORT PEOPLE WITH PAH TO ACCESS THE CARE THAT IS APPROPRIATE FOR THEM

- Encourage shared decision-making between patients and healthcare professionals in the context of multidisciplinary teams
- Provide patients with the information they need to participate in informed, shared decision-making
- Ensure that patients are offered regular risk assessments and are informed by their healthcare professionals about the full range of available, evidence-based treatment options

SUPPORT PEOPLE TO LIVE AS WELL AS POSSIBLE WITH PAH

- Ensure that patients have access to self-management programs to support them to become well-informed patients
- Provide support for caregivers
- Support a holistic approach to care that includes psychological and social support
- Provide patients and caregivers with information on other relevant issues including financial support and disability rights

FURTHER INFORMATION

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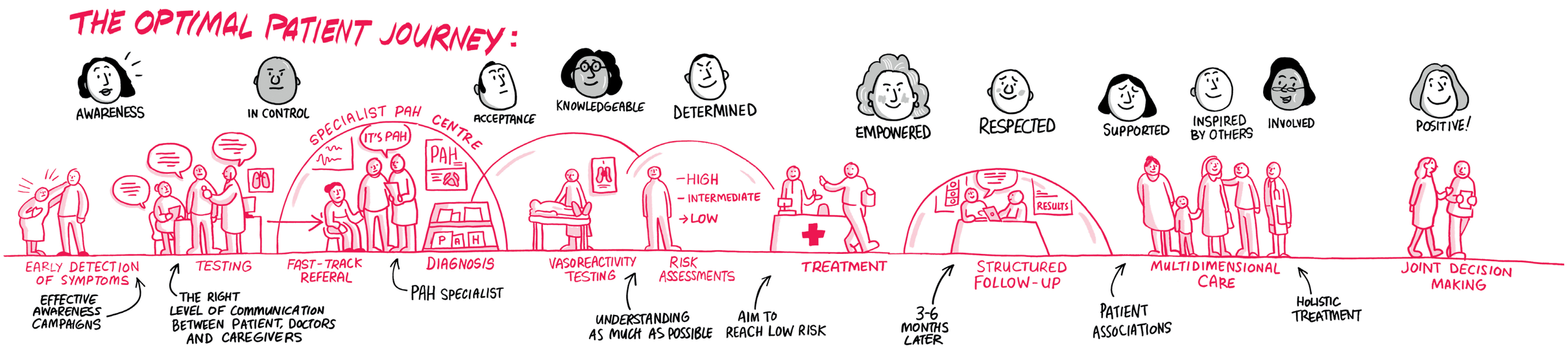

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