



# AMELIORATING CARE FOR PH PATIENTS

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HEQ's editor, Lorna Malkin, speaks to the Project Manager of PHA Europe about the management and treatment of pulmonary hypertension

**P**ulmonary hypertension (PH) is rare pulmonary vascular disease that affects an estimated 25 million people globally. The progressive disease is characterised by a reduction of blood flow from the heart to the lungs caused by the thickening of the arterial walls which can therefore become constricted. This, in turn, puts added pressure on the right side of the heart to keep pushing blood through the arteries of the lungs and can result in the heart becoming weaker and even failing. PH can affect people of all ages and backgrounds but is more commonly diagnosed in women aged between 30 and 60, and those already affected by other heart and lung conditions. Diagnosing the disease as soon as possible is critical since life expectancy can be as little as two years if a patient is not given the necessary treatment required for their specific type of PH. The disease is currently classified into seven subgroups which, importantly, dictates the treatment prescribed to individual patients. Since PH can incur many symptoms of varying severity, it can often be misdiagnosed or even left untreated if diagnosis is delayed. Likewise, despite great strides being taken in the efficacy of treatments available, there is currently no cure for the disease and great disparities in access to treatment across Europe.

Hall Skåra is a Project Manager at the European Pulmonary Hypertension Association (PHA Europe) and part of team working on the so-called 'White Spots' programme which aims to raise awareness of PH by establishing new pulmonary hypertension patient associations in countries where they do not currently exist. HEQ's editor, Lorna Malkin, spoke to Skåra to find out more about the importance of raising awareness of PH across Europe and promoting a more holistic approach to the management and treatment of the disease.

### **To begin, can you tell me what led to the establishment of PHA Europe and the organisation's main aims?**

PHA Europe was established in 2003 in Vienna, Austria. The main aim behind the organisation is that by joining forces, one would be able to make a better impact for the European PH patients. We work for earlier diagnosis, better treatment, better quality of life as well as helping to find a cure. PHA Europe has grown quite quickly, and we now have over 30 associations across Europe.

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Early diagnosis and establishing the correct treatment pathway are really important for people living with PH. PH is multifactorial and can therefore be a very difficult disease to diagnose, often it is mistaken for other conditions. If a person is diagnosed early, they have a longer life expectancy and also a better quality of life. As an organisation, it is important that we can raise awareness of PH and for that reason we are working on multiple awareness campaigns. We work with the pharmaceutical industry because we have a common interest in getting people diagnosed as quickly as possible and ensuring they receive the correct treatments.

Additionally, we have initiated a World Pulmonary Hypertension Day, which takes place on 5 May every year, to advocate greater access to diagnosis and treatment.

### **What are the key symptoms of pulmonary hypertension (PH)?**

One of the key difficulties with diagnosing PH is that the disease can manifest itself in several ways. For most people it simply starts with shortness of breath (dyspnoea) during exertion. Other symptoms can include a racing heart, tiredness and swelling of the stomach, legs and feet. This is why it is so important that we create awareness among the general public and educate them about the symptoms. Similarly, it is essential that healthcare professionals, particularly those first line healthcare professionals like the general practitioner, understand the symptoms because this is who the patient will go to first to discuss their condition. It is really important that the general practitioner is aware that this could be something serious, even though it is most likely that it is not because this is a rare disease.



However, if a young person, for instance, who is mostly healthy and not overweight seeks help as their condition worsens, that should be a red flag to the healthcare professional.

There are many causes of PH, some people can develop the disease without having any other underlying conditions, though it is more likely to occur in people suffering from lung and heart problems including scleroderma, pulmonary veno-occlusive disease and pulmonary capillary hemangiomatosis.

### **How has the COVID-19 pandemic impacted those living with PH in Europe and their access to treatments?**

As PH patients are considered high risk, it is crucial they do not contract COVID and many have isolated themselves even more than the average individual. Some people have been afraid of going to the hospital for check-ups for fear of contracting the infection. Even without the pandemic PH patients need to be careful and do their best to avoid infection. The impact of COVID-19 has varied from country to country. It has not been such a big problem in Norway, where I live, as the pandemic has been pretty much under control. However, in other European countries (especially Eastern Europe), this has been a big problem and particularly because PH patients cannot always access the treatments they need. It is very frustrating to know that you have a very serious chronic disease, and there are medications out there, but you cannot get them and without medication, the life expectancy is 2.8 years after diagnosis.

### **Can you tell me about available treatments and any recent developments or innovations in in this area?**

In the early 90s there were no treatments available for those people living with PH, but in recent years this has really started to change. Nowadays, they have found that if you start to treat this disease aggressively from day one it will give patients a better outcome. We have 12 to 13 effective drugs on the market that will slow the progress of the disease; however, no drugs have been developed so far that will reverse and possibly cure PH.

There are three pathways responsible for cell creation and vasoconstriction and therefore different treatments which target these pathways. Patients may need a combination of medicines to target more than one pathway, depending on how the disease progresses. Other treatments may include blood thinners which can help prevent blood clots, diuretics, and oxygen therapy. If oral medications are ineffective, patients may also be offered IV







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medication which provides continuous medication pumped directly into the heart. If this proves ineffective, surgery such as an arterial septostomy and eventually a lung transplant are the last resort.

I think it is very important to have a holistic approach to treating this disease. People need to learn about diet and how they can exercise safely. Many people get depressed when they become chronically ill, so they may need support from psychiatrists; rehabilitation has also shown to be very effective.

**What key steps would you like to see taken to promote a better quality of life for people living with PH? What could be done at a policy level to improve the availability of treatments and encourage research and innovation?**

Patients need a holistic treatment approach to help them manage the physical and emotional burdens that can arise when living with PH. In addition to taking drugs for their disease, they may need additional support from nutritionists, physiotherapists, psychologists, in order to improve their quality of life. Patient representatives are important during medical trials in order to keep the patient perspective in mind and hence improve the quality of life for people living with PH.

We need to inform the health officials that patients who are treated with the latest drugs will live longer and better and also be able to contribute to society. In this way, they will be an asset to society and not a burden. We are well underway to finding medication that can completely halt the progress of the disease and we will hopefully be able to find a cure if we continue to do research and develop new treatments. It is similar to the situation with HIV patients; before there was no cure, but due to medical research and development, one is now able to keep the disease under control and enjoy a longer and better quality of life.

At PHA Europe, we are working on a call for action paper that will be presented to the EU Parliament. We did one back in 2011 and now we want to present a renewed version because there has been a lot of research and developments in the field. We want to go to Parliament to talk about the issues associated with PH and therefore make healthcare professionals and officials aware of unmet patient needs and treatments.

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