

Many patient journeys, one voice

The recent 'Pulmonary Hypertension Symposium for Patients and their Families' event in Dublin ahead of World Pulmonary Hypertension (HP) Day saw experts in their fields address a packed meeting on how patient groups can use their collective voice to inform health policies and improve the lot of the PH patient overall.

The Seminar, held in the Grand Hotel, Malahide, heard from Prof Seán Gaine, Consultant Respiratory Physician at the Mater Hospital, on 'What is Pulmonary Hypertension and How do we Treat It?'. Prof Gaine was followed by Mr Iain Armstrong, Chairman of the Pulmonary Hypertension Association UK. Mr Armstrong delivered two distinct presentations: 'The Patient in the Middle of the Riddle of PH'; and 'The Journey for the PH Patient and the Role of the Support Group'. Dr Noel McCaffrey, Sports Medicine Physician and Medical Director of BreatheSmart at Dublin City University, delivered a talk on 'The Importance of Exercise in Chronic Illness Rehabilitation' and the symposium was addressed finally by Ms Karen Redmond, Consultant Cardiothoracic Surgeon with the Mater Hospital, who spoke on 'Lung Transplantation for PH'.

Having thanked the attendees for coming — some of whom had travelled considerable distances — Prof Gaine stressed the importance of coming together as a group. "There was a recent example of the power of patient groups with the recent issue of a drug to treat cystic fibrosis," he said. "The power of that group of patients eventually enabled that drug to be available so the voice of patients should not be underestimated."

He also offered his thanks to event organiser Regina Prenderville, explaining that she had recently retired from a previous role, allowing him to opportunistically secure her services. "It really is great to have her as someone who can focus on the work of raising awareness," said Prof Gaine. He also thanked the speakers for attending and provided a brief profile of their respective positions and particularly mentioned Mr Armstrong, a founder of the UK group, as "very supportive and encouraging of what patients, their families and doctors can achieve when they get organised and get together". Dr McCaffrey, he observed runs vital exercise programmes and such therapy is emerging as a new treatment for PH. Ms Redmond, he told the attendees, would provide an important clinical viewpoint as a transplant surgeon with a particular interest in PH.

Different for everyone

"PH is different in everyone," observed Prof Gaine in opening his presentation. "Sometimes it can be the only disorder a person is suffering with, while in other cases, there may be other comorbidities involved." Prof Gaine provided a clinical overview of the nature of PH: "It involves a narrowing of the blood vessels, which are so small as to make them impossible to stent. It is clinically important to note that with Group 1 PH patients, there is narrowing of the vessels in the lung," he continued. "With Group 2 patients, the left side of the heart is affected, slowing down the blood flow through the lungs and this is the most common form of PH seen in Ireland. Group 3 patients have lung disease, while Group 4 patients

commonly experience a clot in the leg, which when it ends up in the lung can block and increase the pressure.” He explained that for this latter cohort of patients, they are most often sent to the UK (Papworth) for treatment because of their experience and outcomes.

“The symptoms of PH can be very vague, which can lead to a delay in diagnosis,” Prof Gaine explained. “The delay to diagnosis can typically be two years, so this is one reason why it is so important to organise events like this to help raise awareness.” He also revealed that for the first time, a database on PH is being established in conjunction with UK (NHS) in order to benchmark the quality of service currently being offered to Irish PH patients with their UK counterparts. “Although generally, Ireland fares as well as other EU countries in terms of outcomes,” said Prof Gaine, who sees between 200 and 300 people at his clinic every year.

Treatment options

Prof Gaine then shared with the attendees an example of an echocardiogram image, pointing out what a clinician sees in a case where the right-heart is enlarged, explaining his system of examination and diagnosis. “The type of treatment then depends on the type of PH the patient is suffering with,” he explained. “Sometimes inhalers are appropriate, whereas other times, drugs to open up the thickened blood vessels are required.” These drugs might include agents to block the production of endothelin, which constricts blood vessels, while other drugs can increase nitric oxide activity. Other agents might reduce prostacyclin production. “Sometimes all three are used in combination, depending what we discover in the range of tests,” said Prof Gaine. In terms of prostacyclin, he observed that nurses “are excellent” at managing any pain that might be associated with this drug, which might possibly be administered via a chronic indwelling catheter.

Looking to the future, Prof Gaine revealed that new drugs are being developed that will change the way PH is managed by clinicians and the patients themselves, and that studies are ongoing to assess the effectiveness of these therapies — and this could be where patient advocacy could play an important role. “Some of the drugs being studied may be shown to have too many side-effects and that would be unacceptable because treating PH is not just about drugs, it is about treating people,” said Prof Gaine, who has worked with the Olympic Council of Ireland. “Athletes are very careful about how they look after themselves and in a way, PH patients can care for their diets and lifestyles with the same attention as these athletes.”

He concluded by looking to the future on a positive note: “The next 10 years will be very important. In laboratories, we can actually cure PH in mice and rats, which have similar biological characteristics to humans. Some day, we may be able to cure PH but we have a lot of work still to do and are looking for markers to predict the development of PH, but the medical profession and patients need to work together.”

Collective voice

Mr Armstrong opened his presentation with a call to PH patients in Ireland to use their experiences to make their collective voice heard. “I passionately believe that patients are an under-used resource,” he said. “The patient is part of

something bigger than the individual and the second part of my talk will address what you can achieve as a patient group.” Presenting slides illustrating the blood vessels in a patient with PH, he continued: “Do we treat PH as a *disease* or an *illness*? This is not semantics, because we can do all kinds of tests for the disease, but the effects of the illness are what the patient has to live with every day.” He described the attendees as “an unusual group”: “This is because each PH patient has to live with their own individual needs.” Mr Armstrong then presented data from the ImPAHct study into pulmonary arterial hypertension (PAH) and its impact on aspects of the sufferer’s quality of life, rather than the clinical disease. “A diagnosis of PH means an uncertain future,” he said. “There is also a financial impact, as 75 per cent of people in this study said the illness had a significant impact on their ability to work. If you have money worries at the same time as having this illness — people might be wondering how they are going to pay their bills, for instance — this can have a major impact on quality of life.” He noted that services for patients with PH had changed rapidly over the past 10-to-20 years and emphasised the individuality of each PH patient. “We all have our own unique life story, our own beliefs and thoughts on our futures,” he said. “PH is most effectively managed with good relations between the patient and the doctor; this is not a ‘quality’ thing, it is just imperative to achieve better outcomes. In some parts of the world, managing PH is just about issuing a prescription, but it much more than that — PH patients can sometimes be overwhelmed by a diagnosis.”

Patients’ voice

With this, Mr Armstrong invited the attendees to consider and reflect on how they felt when they were first diagnosed. “In addition, some employers simply don’t understand what it means when an employee is diagnosed with PH, similarly with insurance companies,” he commented. “Don’t forget the importance of the patients’ voice and the difference that can make to healthcare policies — for example, moves are afoot in the UK to improve the social benefits policy for people with PH and the patient group has been very important in that.” In academic circles, the patient voice is also crucial. “A patient group actually had a paper published in the *British Medical Journal*, which showed that 50 per cent of patients waited 11 months on average before going to their GP. They often just held on to their symptoms and it was often the husband or wife who said something like, ‘Something is not right with you’.”

The challenge, said Mr Armstrong, was for medical professionals to recognise the symptoms early, as he noted that 50 per cent of PH patients had to see four or more doctors before a diagnosis was made, a situation he described as “unforgiveable”.

“Quality of life means different things to different people,” he said. “Specialist care can be expensive and patients often have to travel some distance to get it. On average, there can be two days of specialist care and the other 363 days are self-management by the patient and it’s important for you to spot it when the symptoms change.” Again emphasising the individual rather than the clinical disease, Mr Armstrong quoted Sir William Osler, who once said: “Ask not what disease the person has, but rather what *person* the *disease* has.”

Oxygen use

His presentation was followed by a lively and informative Q&A session. One attendee — who's diagnosis took two years — asked the panel why the post of 'Consultant General Physician' had disappeared in recent decades. Prof Gaine suggested that there may be a move back in this direction in coming years with the post of acute medicine physician. Another patient enquired as to the use of oxygen in symptom control. Prof Gaine replied that for some people, oxygen was crucial while for others, it provided a certain sense of comfort, but the question was hard to answer because of the various different types of PH. Mr Armstrong added that according to a survey in the UK, oxygen could sometimes make people feel isolated and they may not "be bothered with the hassle" when they were going on holiday, for example.

Dr McCaffrey's presentation was preceded by Prof Gaine revealing that in approximately six months, guidelines will be published that indicate exercise as a form of therapy in PH.

Exercise and PH

Dr McCaffrey began by stating that there is not a single illness where exercise cannot play a therapeutic role. He also stated his own special interest in the role of a patient group in pushing the PH agenda: "It's extremely important for a patient group to have ownership of the issue and help to design treatment policy," he told the meeting. "One of the benefits of engaging in an exercise programme is that it limits isolation because people go to their exercise classes and meet many other interesting people."

Dr McCaffrey elaborated on the role of exercise in disease treatment and prevention: "Some cancers are driven by fat content," he explained, "and exercise can also help to counteract cognitive decline. In fact, there is only one cancer that is common in people who exercise a lot — skin cancer."

Exercise can also greatly improve quality of life, said Dr McCaffrey, "If you have reduced strength, you will have trouble doing certain tasks, such as getting up, using the toilet and so on. If you have reduced flexibility, you may have trouble reaching things and bending over." Dr McCaffrey presented slides showing that no matter what age, rate of physical and cognitive decline can be slowed by 1,000 per cent in a structured exercise programme.

His clinic in DCU deals with people suffering with chronic organ failure, social issues, psychology issues and even incorporates spiritual aspects. "There is a Chaplain getting involved soon, so that's great," said Dr McCaffrey. He presented examples of chronic disease patients mixing with elite athletes — such as Dublin footballer James McCarthy — in his clinic, and emphasised: "Sometimes people say, 'Doc, I'm not getting any better', but in fact halting a decline in a disease can in itself be a very positive outcome. One of the things that stop PH patients exercising is that if their lungs are incapable of supporting five minutes of exercise, this can become frustrating for them. So we break that down into five one-minute blasts to make it less frustrating for the patient."

Aerobic capacity

It has also been shown that even doing strength exercises alone can improve aerobic capacity, said Dr McCaffrey, because the harder muscles work, the greater the increase in lactate capacity. "Using oxygen can also make it easier to

exercise,” he explained, presenting data (Grunig et al, 2012) showing huge improvements in quality of life across a range of patient groups.

“The most common questions we are asked are, ‘What can we do about my sore knee, sore back, sore hand’, and so on. Remember that any of these can normally be helped by exercise. If there is an improvement in quality of life, strength and flexibility, then that is a small price to pay for an evening of stiffness,” he concluded.

Humble beginnings

Mr Armstrong opened his second presentation, which focused on what a patient group can do to champion improved services, although he was keen to give a truncated talk to allow for more questions from patients. “PHA UK had humble beginnings,” he told the room. “It was almost like a peer support group of about half a dozen people. They just got together to talk about the practicalities of everyday life. Now the organisation has 2,500 members and is a great forum to make things happen. The real benefit is to link people up and curb isolation, which can even occur in the family setting.”

Addressing the patient journey, he observed that for most people the diagnosis is the first part of this. “Often patients and their families distrust medicine so the initial six months is usually spent getting people on board and convincing them that we can help. The second part is administering medical treatment,” he said. “I like subcutaneous treatment because when it works, it works really well but the biggest change in PH treatment in the past 15 years has been to move from treating PH as an acute illness to a chronic condition, which puts exercise on the agenda, as nobody would have suggested exercise as a form of treatment for an acute illness.”

He called on the attendees to use their lobbying power: “As a patient organisation, you have to understand the journey, you need to produce resources such as booklets and websites. World PH Day is approaching and we have to increase awareness and develop a strategy to ensure your voice is heard.”

Mr Armstrong concluded: “Treatments are constantly changing but we have a fight on our hands to get access to these drugs, but don’t underestimate the power of the patient voice — talk to the politicians; they are fragile in some respects and don’t like bad publicity because it affects the ballot box.”

A second Q&A session followed, with one attendee asking why all treatment pathways were not used all together in PH. Prof Gaine replied that there were three specific, high-tech drugs but using all of these together could produce too many side-effects and make the patient’s quality of life worse. Therefore, they were introduced sequentially and a determination made about the need for a second agent if the progress wasn’t satisfactory. The next five years should shed more light on this, he replied.

Another PH patient asked whether personalised medicine could play a role in PH treatment. Prof Gaine responded that this was a “new frontier” because different people respond to different drugs. Another attendee asked whether there was any financial help available for patients who need to use oxygen 24/7, in light of the impact this has on electricity costs. The answer was that unfortunately, no

aid was available to Irish PH sufferers but in the UK, patients could receive a 10 per cent discount on their electricity bills.

Transplants in PH

The final presentation was delivered by Ms Karen Redmond, who opened by declaring her special interest in PH and outlined new techniques in transplantation. "Organs are a bit like engines; sometimes they just need to be tweaked and sometimes they just have to be replaced," she told the room. "There has been a lot of work in the area of lung transplants (LT) in the last few years, despite very little money going into LTs in Ireland. The National Transplant Advisory Group has put a business plan to the HSE so we will see what happens there."

The transplantation process begins with "the gift of donation, which is a gift of life," said Ms Redmond. "Often, a grieving family can take great comfort from knowing that they have done this and this is reinforced when they sometimes receive thank-you letters from the recipient."

In organisational terms, she explained: "The Intensive Care Society has donor management guidelines but we need dedicated link nurses who have accountability; at the moment, people do this in their free time, but this needs to be a more formal role. Irish people are very generous as donors and the amount of offers we get is very high, relative to the amount of funding and support we receive."

Ethics and procedures were also important, she stressed. "The National Ethics Committee outlines different donor circumstances and guidelines on non-brain-stem death. Some people who are in an Intensive Care Unit will never realistically make it out of there, for example. We also need to have some quality control so that we can benchmark ourselves against other countries," said Ms Redmond.

"With the EU Directive, hopefully we can organ-share and there may be an opportunity with Northern Ireland and the UK and others, where we could become the 'net importer' instead of the 'net exporter' of organs."

Strength and body weight

Transplants are only necessary in a small amount of patients, she explained, and there can be contraindications in patients who have cancer or cardiovascular disease. In PH patients, this can also be the case because of comorbidities. "I can give people pristine lungs, but strength and body weight are important for survival," she commented. Ms Redmond then presented a range of slides and talked through the donation and transplant procedure. "There is a discussion that takes place on the night with the medical team and recipient," she said.

"Irish people sometimes have the idea that they should be on as little oxygen as possible but oxygen is nutrition for the lungs. When the lungs are replaced, the right side of the heart recovers remarkably quickly and people do much better after transplantation if they comply with their medication."

On treatment options, Ms Redmond said that some patients are given a small hole in the right-heart to relieve pressure and this is not routinely closed during the operation because this can increase surgical stress.

"Transplant offers a better quality of life and people can live longer but we can run into problem with rejection or infection," said Ms Redmond.

“We would love to have a crystal ball to know how long someone will be on a transplant list and how long someone will get from their native organs.” She outlined the profile of the “perfect donor”; a non-smoker, less than 55 years old, with a clear bronchoscopy. “One of the initiatives we have introduced is the idea of extended or ‘marginal’ donors, where we can use lobes instead of the whole lung,” she explained. “We sometimes put a patient on a bypass machine for the operation because when they come for transplant, their PH will be at a very advanced stage.”

To conclude, Ms Redmond then presented a case study of a 19-year-old cystic fibrosis patient, including x-rays and a video of the perfusion, and outlined the peri-operative supports in place, which involved a “very cautious and safe approach”.

Exercise prescription

In the final Q&A session, one patient expressed frustration that she could not find an exercise programme that would take her on. Prof Gaine revealed that work will be done on preparing a prescription that would allow people to access these programmes, and that this would hopefully happen within the next year. Another attendee asked about the possibility of a sponsorship programme, or mentor scheme, for PH patients who were misdiagnosed. Prof Gaine responded that it may be difficult to “link people up” because of confidentiality issues, and this was another strong reason for the existence of the support group.

A number of the attendees expressed their gratitude to the speakers and organisers and stressed how useful the Symposium had been for them and their families, with the exercise programme generating a lot of interest.

Poignantly, one interesting aspect of the Symposium involved four empty chairs; rather than taking their seats on the stage behind a table, all the speakers rather opted to sit with the PH patients and their families as they listened to each other’s presentations and fielded questions.

This epitomised the spirit of the event — patients, their families and medical professionals together in the drive to improve the lives of those with PH.