

Recommending Exercise in Pulmonary Hypertension: Adult and Pediatric Perspectives

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Catherine Avitabile and Aimee Layton asked questions of their colleagues from around the world on their experiences with exercise recommendations in PH patients of varying ages. They discussed patients' and their families' views on physical activity, benefits and risks of engaging in exercise, and resources for both health care providers and patients.

Avitabile/Layton: Do you find most of your patients exercise regularly or are physically active when you talk to them about their daily routines?

Chia: Most are quite sedentary.

Benjamin/Grünig: Patients with pulmonary hypertension are often very active or used to be very active before the onset of severe symptoms and their initial diagnosis. This changes with the recommendation to avoid physical exertion and an uncertainty how much exercise can be performed without causing harm and worsening the disease. Consequently, patients with pulmonary hypertension are often highly motivated to regain physical activity in their daily life.

Berman Rosenzweig: I find that this is quite variable between patients. Many patients are highly motivated to exercise once they feel up to it. I believe, in addition to helping their overall physical fitness, there is an important sense of empowerment that can't be quantified but I see all the time. Being able to exercise and know that you are contributing to your wellness in PH can make a real difference for patients. For those that are not feeling well enough to exercise, we

still try to encourage them to participate in cardiopulmonary rehab to try to enable reconditioning.

Avitabile/Layton: How do patients/families react to a discussion around physical activity?

Chia: People are very open to it once they've had some discussion/education on benefits and safety. The main concerns are whether exercise is safe for them.

Benjamin/Grünig: Patients who plan to enter a rehabilitation program often seem to have been waiting for specialized training and an answer to the question which exercises can be done safely. They are keen to be active again and motivated to learn techniques to improve their exercise capacity. Patients' families are usually supportive and very much appreciated, when the exercise capacity improves and daily activities are enabled, e.g., going for a walk together. The prospect of this improvement is motivating for both patients and families.

Berman Rosenzweig: Patients and families often have many questions about physical activities and the dos and don'ts. This is one of my favorite conversations because one of the many goals of treatment is to enable patients to be able to resume their usual activities which includes exercise. There are precautions of course, for example, no heavy weightlifting, but we review these in our conversations together.

Avitabile/Layton: What do you perceive as the benefits of routine physical activity or planned regular exercise in your patients?

Chia: There are many, such as increased exercise capacity and therefore functional capacity, peripheral physiological adaptations (increased capillaries in muscle, increased muscle strength), and possibly central adaptations. It can also contribute to increased quality of life and engagement/agency.

Benjamin/Grünig: In our studies, we have already shown that a specialized exercise training program can improve exercise capacity, with improvement of 6-minute walking distance and peak oxygen consumption, quality of life, symptoms, and possibly right heart function. In addition, patients learn which exercises they can do to support their well-being and to be active again. In our program, patients also learn to avoid overexertion, which is key to safe exercising in PH. Whether this also has an impact on the clinical course and stability of the disease still needs to be investigated.

Berman Rosenzweig: There are many benefits to exercise, both physical and emotional. Patients often don't realize how deconditioned they've become when they weren't feeling well even before they were diagnosed. They often self-limit due to their breathing limitations, and this leads to physical deconditioning. So there is a real benefit, once feeling well, to reestablishing regular exercise. Muscle reconditioning is important, but being in better shape overall is less taxing on their heart and lungs when they exert themselves.

Avitabile/Layton: Are there specific guidelines you follow when recommending planned exercise to your patients?

Chia: No specific guidelines—more general safety recommendations (e.g., Borg RPE, symptoms to watch out for). Anything more strenuous than gentle walking I would usually recommend starting exercise in a supervised manner, e.g., cardiac or pulmonary rehab program.

Benjamin/Grünig: We do not recommend patients perform exercise training on their own. Patients often tend to overexert themselves. Therefore, exercise training should be taught in a supervised setting and consecutively transferred to the patients' daily routine. Our exercise training program was specifically developed for patients with pulmonary hypertension. In contrast to other training programs, we use a low-dose training intensity. However, it has also been shown in professional athletes that modern exercise programs are mainly focused on lower intensity training which is highly effective if it is individually adjusted. A key characteristic of our program is also a close supervision and adaptation of the exercises to the individual patient. This is enhanced by assessment of heart rate, oxygen saturation, subjective estimation of exertion, and information on right ventricular function throughout different stages of workload during an entrance examination.

Berman Rosenzweig: We work very closely with our exercise physiologists or with local cardiopulmonary rehab programs to establish guidance for our patients. Again, each patient is different and will have different constraints so the exercise guidance must be individualized.

Avitabile/Layton: What resources do you access when making these recommendations?

Chia: For patients, we use plain language guides, like those on PH Websites about exercise and PH. For myself, I access evidence-based research articles.

Benjamin/Grünig: The specialized training in Heidelberg is started as an

inpatient rehabilitation for 3 weeks in cooperation with the rehabilitation clinic Koenigstuhl. At the beginning and at the end of the in-hospital phase, a detailed examination gives insight into the patient's exercise capacity, individual goals, and challenges. After continuation of the program at home for 3 months, a follow-up examination is offered to the patients to have feedback on the training effects and to discuss training adaptation.

Avitabile/Layton: What barriers do you face when recommending increased physical activity or planned exercise in your patients?

Chia: Personal factors such as fear/anxiety, breathlessness, lack of time, and lack of support/motivation. There are also logistical factors like access to appropriately supervised exercise, cost, or weather.

Benjamin/Grünig: In Germany, the health insurance companies mostly cover the costs for a rehabilitation program. However, it is not always easy to receive cost coverage for the specialized PH program, as this is more individualized and has higher costs. Insurance companies are often also attached to certain rehabilitation clinics, which makes it harder to receive cost coverage for a specific clinic and program.

Due to the inherent risks of exercise training in patients with right heart failure and to avoid worsening of the disease or an ineffective training, we recommend patients to take part in a specialized program rather than a general program to enhance physical activity. The specialized program in Heidelberg was specifically developed for patients with pulmonary hypertension and was continuously scientifically evaluated. Learning the right exercises and techniques is key to safe and successful training in these patients and should not be initiated in a general recommendation to be more active. If the exercise training is too intense, it can lead to

disease worsening and increasing right heart failure.

Berman Rosenzweig: Patients can be apprehensive. In these cases, I do think it's important to work with rehab programs that can help monitor during initial trials of exercise and give tangible parameters (oxygen saturation and heart rate, for example) so patients can self-regulate when comfortable exercising on their own.

Avitabile/Layton: What type of research do you feel would be impactful in this area?

Chia: I think it's clear that exercise improves exercise capacity and QOL in patients with PH. I would love to see more studies about possible central benefits (e.g., effect of exercise on SV, CO, mPAP, etc.). However, these studies are invasive and difficult to recruit.

Benjamin/Grünig: We are still in need of more research in this area, though the impact of exercise in pulmonary hypertension has already been investigated in numerous studies. Especially the impact on the long-term outcome, time to clinical worsening and survival, right heart function and the mechanisms of action have still to be investigated. Furthermore, there is a lack of data on the optimal training methods and setting in these patients, and of course, training effects on different types of pulmonary hypertension as well as stages of severity should be focused on in future research.

Berman Rosenzweig: I believe we are just on the early cusp of integrating wearable technology into our day-to-day practices. Measures such as sedentary time can be so informative about the day-to-day activities or limitations our patients face. Heart rate trends and other measures in real time can also tell a much richer story about the real daily activities our patients can achieve and help us to tailor treatment. I would really love to see more actigraphy studies integrated into clinical trials for PH.