PH PROFESSIONAL NETWORK "Just Do It": Practical Aspects of Pulmonary Rehabilitation Programs

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WHAT IS PULMONARY REHABILITATION?

Pulmonary rehabilitation (PR) is a specialized physical therapy program to improve the quality of life for patients who have symptoms from chronic lung disease. The American Thoracic Society offers a formal definition of PR as a:

... comprehensive intervention based on a thorough patient assessment followed by patient-tailored therapies that include, but are not limited to, exercise training, education, and behavior change, designed to improve the physical and psychological condition of people with chronic respiratory disease and to promote the long-term adherence to health-enhancing behaviors.¹

Although PR was originally developed to address the needs of patients with the most common forms of lung disease (which result from smoking),² the goals of PR can be applied to help patients with other types of lung disease, including patients with pulmonary hypertension.³

WHAT ARE THE IMPORTANT COMPONENTS OF A PULMONARY REHABILITATION PROGRAM?

While a significant portion of the program is devoted to exercise sessions

for improving physical conditioning, PR provides an opportunity to optimize patients' self-management skills to reduce the frequency and severity of pulmonary symptoms, reduce the risk for hospitalization, and help address anxiety and depression that may result from their chronic lung disease. This helps patients to engage in daily activities, participate in events with family and friends, and return to their previous level of function after a hospitalization or recovery from an exacerbation of their pulmonary condition. The benefits from PR are not just from patient contact with the medical staff, but also from supportive interactions with other patients enrolled in the program.

Pulmonary rehabilitation programs are staffed by a multidisciplinary team. This is important for tailoring the program to a patient's specific needs. The core members of a PR program often include doctors, nurses, and respiratory therapists. Most programs will also integrate additional staff, including exercise physiologists, physical and occupational therapists, nutritionists or dieticians, and psychologists or social workers.

Although regular visits to doctors remain important to define a patient's medical treatment plan, PR offers a supportive environment to "fill the gap" and empower patients with skill and confidence needed for managing their chronic lung disease. After an initial visit, an individualized treatment plan is created by input from the multidisciplinary team and tailored to the patient's specific needs. Some of the common topics addressed by PR include:

- Education to improve the patient's understanding of their chronic lung disease;
- Review breathing techniques and energy conservation strategies;
- Review techniques and devices to support airway mucus clearance;
- Address questions regarding medications for maintaining stable chronic lung disease;
- Review how and when to use home oxygen (O₂) therapy and other respiratory devices;
- How to recognize symptoms that indicate deterioration of his or her chronic lung disease;
- Identify and reduce the impact of depression, anxiety, and other psychosocial stressors that are more common among patients with chronic lung disease;
- Review and address questions about the action plan recommended by the patient's local doctors.

WHY IS PULMONARY REHABILITATION USEFUL?

When patients start to experience symptoms from chronic lung disease, there is a natural tendency to hold back or slow down because they do not feel well, avoiding activities that cause shortness

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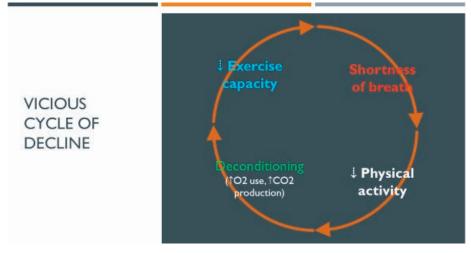


Figure 1: Patients who experience symptoms of chronic lung disease, such as pulmonary hypertension, will often refrain from physical activity. When patients become less active and stop exercising, this leads to deconditioning, meaning that their muscles become less efficient, and this places even more demands on their breathing. This results in a negative impact on their exercise capacity, and they will experience shortness of breath at even lower workloads.

of breath. When patients become less active, this leads to deconditioning, meaning that their muscles become less efficient, using more O_2 and producing more carbon dioxide (CO₂) during any activity, and this places even more demands on their breathing.

As patients become more deconditioned, they will experience shortness of breath at even lower workloads, creating a vicious downward spiral of deterioration (Figure 1). Additionally, how patients respond to stress and discomfort will also influence their behavior, meaning that some patients become anxious when they are short of breath, and this causes them to breathe faster than necessary and adds even more to their shortness of breath.

Through monitored exercise sessions that are tailored to a patient's specific limitations and needs, PR can help patients to overcome deconditioning, reducing the demands that physical activity imposes on their heart and lungs. With improvements in exercise tolerance and reduced shortness of breath in response to physical activity, many patients will feel more confident about re-engaging with friends, family, and other activities they may have avoided or abandoned when they were not feeling well. In turn, PR can help patients be less worried and distracted by their lung disease and function better in the real world.

ARE THERE ANY SPECIAL CONCERNS FOR PATIENTS WITH PULMONARY HYPERTENSION ?

Patients with PH have special reasons for impaired exercise capacity. Although different patients may have different reasons for having PH, the main symptoms and problems that patients have are due to increased blood pressure in the lungs (the definition of PH). On average, a person's total blood volume is approximately 1 gallon, and it is the heart's job to pump blood out

LUNG AND HEART

IMPACT EXERCISE

FACTORS THAT

CAPACITY

from the left ventricle to the other major organs through the arteries. The blood then returns back to the heart through veins, and it is the job of the right ventricle to pump the same 1 gallon of blood per minute through the lungs.

For patients with PH, the right ventricle must work harder to pump blood through the lungs, and when they engage in physical activity, either through exercise or just from their routine daily activities, this increases the demand to deliver more O_2 to the muscles of the arms and legs and a need for the lungs to get rid of additional CO₂ being produced by the muscles (Figure $\overline{2}$). Pulmonary hypertension will also cause some patients to have low O₂ levels, both at rest and during physical activity, causing the heart to work even harder to pump more blood with less O_2 in it to support the muscles and other major organs. As a patient's PH worsens, the right ventricle might not be able to keep up with this work load, causing shortness of breath, leg swelling, and early fatigue during physical activity.

If a patient's heart cannot keep up with the increased workload caused by PH, then patients will start to show signs of right heart failure, which means that the right heart is unable to pump enough blood through the lungs to meet the needs of other major organs, causing other symptoms including:

- Deconditioning = Muscles become less efficient
 T O2 use, ↑ CO2 production
- Deconditioning + Loss of muscle $\rightarrow \frac{1}{2}$ Exercise capacity
- Effects of pulmonary hypertension during exercise
 - \uparrow Blood pressure in the lung $\rightarrow \uparrow$ Load on right ventricle of the heart \rightarrow Heart must work harder \rightarrow Cannot maintain O_2 delivery to muscles \rightarrow Fatigue and shortness of breath
- Reduced oxygen levels
- Effects of lung disease during exercise
- Lung disease → Unable to breathe more deeply → Must breathe faster
 - Weakness of breathing muscles
- Obesity → Must work harder to breathe

Figure 2: In addition to the negative effects from deconditioning, pulmonary hypertension will impose an additional load on the right ventricle on the heart, exacerbating the degree of fatigue and shortness of breath during physical activity. These problems are made worse by coexisting lung disease, which magnifies the work of breathing needed to support the metabolic demands from exercise.

- Hypotension
- Tachycardia
- Syncope and pre-syncope
- Reduced urine output and other signs of kidney damage
- Nausea or abdominal pain

For these reasons, it is important for PH providers to assess and document that the patient's PH is stable and they do not have signs of right heart failure before starting an exercise program. This is also why it is often useful for PH patients to begin to exercise under the supervision of a PR program. The PR staff will routinely monitor patients for signs of hypotension, tachycardia, hypoxia, chest pain, or syncope. Many standard rehabilitation orders include the administration of nitroglycerin for chest pain. This order should be removed for PH patients receiving sildenafil, tadalafil or riociguat due to a drug interaction.

For those patients who have PH that is caused by another lung disease, such as emphysema or pulmonary fibrosis, this creates even more difficult problems. When patients with both PH and another lung disease experience shortness of breath, they are unable to simply take deeper breaths; they can only breathe faster when short of breath or when trying to exercise. By breathing faster than normal even during low level physical activity, this causes their breathing muscles to need more O_2 and produce more CO_2 , which in turn places more demands on their lungs and breathing muscles.

Patients with PH due to another lung disease often have great difficulty maintaining normal O_2 levels and require very high flow O_2 during physical activity. This is often another reason for patients to start exercising under the supervision of a PR program where very high flow O_2 is available. The combined effects of deconditioning, PH, and lung disease cause patients to have very limited exercise capacity, and sometimes a modified exercise program using interval training should be considered until the patient can tolerate longer intervals of physical activity.

Pulmonary rehabilitation programs also offer an opportunity for PH patients to meet with a nutritionist to address individual needs, including reviewing low sodium options for patients with leg edema or other signs of fluid overload, diet modifications for weight control, and review nutritional strategies to preserve muscle mass for patients who have very severe lung disease.

WHO CAN ENROLL IN PULMONARY REHABILITATION?

Government regulations established by Centers for Medicare & Medicaid Services outline how PR for patients with chronic obstructive pulmonary disease (COPD) is supported by health care coverage. These guidelines outline how PR is covered by Medicare for patients with moderate to very severe COPD. However, coverage for other lung problems is determined on a caseby-case basis, and coverage may differ depending on insurance plans. For cases where insurance coverage is limited, it may be helpful to have a discussion with the PR program about the appropriate diagnosis and billing codes that could be used to maximize reimbursement. The most common lung problems addressed through PR include:

- COPD (which includes emphysema and chronic bronchitis);
- Interstitial lung disease (including idiopathic pulmonary fibrosis);
- Sarcoidosis;
- Pulmonary hypertension;
- Bronchiectasis;
- Lung cancer and lung cancer surgery;
- Lung volume reduction surgery;
- Before and after surgery for lung cancer or lung transplantation.

Referrals to the PR program must be made by a signed physician order. This must be accompanied by medical records from the past 6 to 12 months that include physician notes, chest x-ray or computed tomography (CT) scan report, pulmonary function tests, electrocardiogram, and bloodwork results. Additional details regarding a patient's PH may also be requested, such as reports from echocardiogram and right heart catheterization.

For patients with certain medical problems, such as diabetes, a special

action plan may need to be discussed with the patient's primary doctor. Exercise training is a healthy approach to lowering blood sugar levels. Patients with diabetes may be asked to check their blood sugars before and after each PR session, and it will be necessary for the patient to review this information with their primary doctor first to see if they should change how they take their diabetes medications on the days they attend PR.

For patients who do not have access to local PR programs, cardiac rehabilitation programs or supervised exercise by physical therapy programs can be considered if insurance coverage is available.

OPTIMIZING USE OF PULMONARY REHABILITATION Limited Awareness

Pulmonary rehabilitation has proven to be an important and safe treatment option for patients with chronic lung disease. Even for patients with severe PH, successful completion of PR can improve a patient's exercise capacity and quality of life, reduce shortness of breath and fatigue, and possibly even improve the management of their PH.⁴ Although PR is considered to be part of the standard of care for patients with symptomatic chronic lung disease, medical authorities are concerned that many patients who would qualify and benefit from PR are not receiving PR as part of their PH management plan.⁵ Raising awareness in patients and their doctors of the availability of PR would be one way to maximize the benefits to patients with PH.

Health Care Coverage Limits

Similar to how many dental insurance plans set a lifetime limit for orthodontic treatment (for example, braces), some health insurance plans set a lifetime limit for PR coverage, even though, from a patient's perspective, they may need PR for what is usually a lifelong condition, such as PH or COPD. For patients who experience frequent hospitalizations related to their lung disease and have a lifetime limit for PR coverage, the patient must review alternative approaches with their primary provider and PR program, including transitioning to a phase 3 pulmonary maintenance program (PMP), described below.

Pulmonary Maintenance Program

It remains important for patients to continue to exercise routinely and maintain health-enhancing activities (low sodium diet, proper use of O₂, refraining from tobacco use) gained through the standard outpatient phase 2 PR program. To assist with this, many PR programs offer a phase 3 PMP. This allows patients continued access to the PR equipment and staff at a lower cost, particularly if they are unable to make arrangements either on their own or through a local exercise facility. Pulmonary maintenance programs are usually intended for recent "graduates" from a standard phase 2 PR program, where the patient is familiar with the exercise procedures and equipment and they have been observed long enough to confirm that they are motivated and capable to exercise safely with less direct supervision.

Logistics of Pulmonary Rehabilitation

Since the inauguration of the first PR programs, patients referred to PR, on average, have become older and have

more medical problems. This imposes new challenges to successful participation in PR. Of course, doctors should take care to help patients enroll in a program closest to their home or place of work; the American Association for Cardiovascular and Pulmonary Rehabilitation maintains a directory: https:// www.aacvpr.org/Resources/Resources-for-Patients/Pulmonary-Rehab-Patient-Resources.

To ensure that patients can maximize the benefits gained from PR, patients are encouraged to participate at least 2 days per week. Many patients have additional demands on their time and may need to make time for other medical visits, family events, planned trips, and unplanned acute illness. One specific example: a patient who requires outpatient dialysis must make sure that their dialysis schedule is compatible with the available PR sessions. Patients who rely on public transportation, mobility services, or require additional assistance to bring them to PR must take care to make arrangements in advance.

Formal PR as an adjunct therapy for PH is now embedded in the guidelines for standard PH care. To successfully enroll and complete PR, careful collaboration between the PH patient, PH providers, and the PR program is required for safety, efficacy, adherence, and insurance coverage.

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