PF is different from chronic obstructive pulmonary disease (COPD) and cystic fibrosis (CF)

There are many important differences between PF and other forms of chronic lung disease, including symptoms, rehabilitation strategies, and oxygen requirements.

Characteristics	PF	COPD	CF
Cough	Usually dry	May be dry or productive	Moist with purulent sputum
Chest Physical Therapy	Ineffective	Infrequently required	Required daily or more often
Exertional Oxygen Requirement	Often much higher than resting requirement	Exertional oxygen require-ments usually modest	Varied
Pulmonary Rehabilitation	Recommended; Refer early in the disease course if possible	Recom-mended	Not recommended
Exercise Training	Aerobic and resistance training; Consider strategies to minimize excessive desatu-ration if this limits the exercise dose	Aerobic and resistance training	Aerobic and resistance training

Resources

Adventures of an Oxy-Phile2, Thomas L. Petty, MD

ATS/ERS Statement: Key Concepts and Advances in Pulmonary Rehabilitation in Am J Respir Crit Care Med. 2013 Oct 15;188(8):e13-64. doi: 10.1164/rccm.201309-1634ST

SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION

The mission of the Pulmonary Fibrosis Foundation is to accelerate the development of new treatments and ultimately a cure for pulmonary fibrosis. Until this goal is achieved, the PFF is committed to advancing improved care of patients with PF and providing unequaled support and education resources for patients, caregivers, family members, and health care providers.

To learn more about how the PFF can help support you, contact the PFF Help Center at **844.TalkPFF** (844.825.5733) or **help@pulmonaryfibrosis.org,** or visit the PFF online at **pulmonaryfibrosis.org**.

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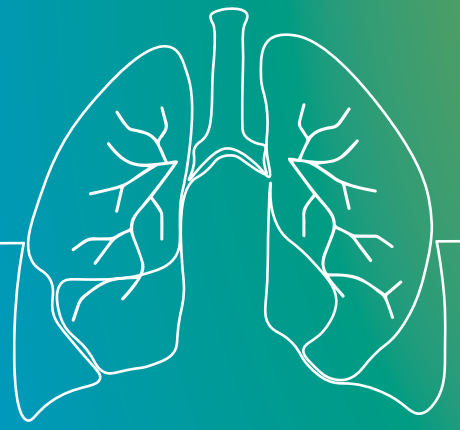

ABOUT PULMONARY REHABILITATION

A GUIDE FOR ALLIED HEALTH PROFESSIONALS

Pulmonary fibrosis (PF) refers to all of the fibrotic interstitial lung diseases (ILD), including idiopathic pulmonary fibrosis, chronic hypersensitivity pneumonitis, pulmonary sarcoidosis, autoimmune-related ILDs, and others. People living with PF frequently experience reduced exercise capacity, breathlessness on exertion, cough and fatigue that limits daily activities.

Respiratory therapists and physical therapists are key members of the multidisciplinary team that cares for people with PF across the course of the disease, and may provide a number of interventions to manage symptoms, enhance functional exercise capacity and improve wellbeing. This brochure outlines some treatment strategies that may be considered for people with PF.

PULMONARY REHABILITATION

There is good evidence for the short-term benefits of pulmonary rehabilitation (PR) for people with PF, including improved quality of life and better exercise tolerance, and this treatment is recommended in clinical practice guidelines. The benefits may be more long lasting if pulmonary rehabilitation is undertaken earlier in the disease course, so early referral should be encouraged. Pulmonary rehabilitation programs should consist of supervised endurance and resistance exercise training, disease education and support.

During exercise training some patients may require strategies to prevent excessive oxyhemoglobin desaturation, for example oxygen supplementation, interval training, downhill walking or resistance training. Patients with clinically significant pulmonary hypertension should undertake an exercise training program under the supervision of an experienced health professional. All patients who undertake pulmonary rehabilitation should have a thorough assessment prior to commencement, including (at minimum) a validated measure of exercise capacity (e.g. 6-minute walk test) and quality of life. These measures should be repeated at the end of the program to quantify any changes and guide future therapy.

Patients who are unable to access a PR facility, or would like to continue an exercise regimen following the conclusion of an in-person program, may benefit from the PFF's Pulmonary Rehabilitation Toolkit (pulmonaryfibrosis.org/PRToolkit). This program, created in partnership with the American Association for Cardiovascular and Respiratory Care (AACVPR), consists of eight self-paced modules and includes exercise videos, reading material, downloadable tools for tracking progress and vital signs, and mindfulness resources. Patients should receive approval from their physician before utilizing the PR Toolkit.

PHYSICAL ACTIVITY

Being physically active is associated with better health outcomes in PF. People with PF should be encouraged to increase or maintain their physical activity levels, consistent with general population guidelines of 30 minutes of physical activity on most days of the week, at a moderate intensity equivalent to a Borg dyspnea score of 3-4 on a 1-10 scale. Physical activity can be accumulated in shorter bouts of 10 minutes at a time if needed.

OXYGEN

Many patients with pulmonary fibrosis require long term oxygen therapy and should be encouraged to use their oxygen therapy as prescribed with rest, during exercise, sleep and any changes in altitude. Patients using oxygen may require higher flow rates during exercise. Many patients with PF exhibit profound exertional desaturation. If the patient has been prescribed portable oxygen only, they should be educated on its use during exercise, rather than just during recovery. Selection of the appropriate device for ambulatory oxygen is critical, taking into account the patient's oxygen requirements, mobility, their environment and ability to manage the device independently.

MANAGING SYMPTOMS - DYSPNEA, FATIGUE, PAIN, COUGH

Many people with PF achieve improvements in dyspnea and fatigue with pulmonary rehabilitation. Pacing and energy conservation strategies can be useful as patients may need to slow down and take rests, to maintain physical activity levels and participate in activities of daily living without intolerable dyspnea or fatigue. Pulmonary fibrosis is often characterised by a distressing, dry cough that is difficult to control and significantly impacts quality of life. At present there are few evidence-based strategies to reduce cough in PF, but there are several agents being investigated in clinical trials (PFF Clinical Trial Finder). Traditional chest physical therapy techniques are not generally effective. Some patients report benefit from remedies such as cough lozenges (especially those containing an anaesthetic agent), sipping water or breathing strategies such as pursed lip breathing. Please discuss with the physician regarding whether prescription of cough suppressants is indicated.

MANAGING COMORBIDITIES

Musculoskeletal pain is common and may interfere with daily life activities and exercise participation. Appropriate assessment and treatment from a skilled musculoskeletal physical therapist should be considered. Depression and anxiety are also very common and patients should be referred for appropriate management.