Symptom Control for Patients with Interstitial Lung Disease
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- The dyspnea experienced by patients with ILD is multifactorial and correlates more strongly with quality of life than with pulmonary function parameters. Pulmonary Rehabilitation (PR) programs provide physical conditioning, education, and social support and should be considered first-line treatment for the dyspnea, fatigue, depression and anxiety that often co-exist in ILD patients. Recent awareness of the need for more ILD-specific PR content will boost the PR benefit for these patients.

- The understanding of the mechanisms of cough in ILD is advancing and includes the interplay of micro-aspiration, upregulation of airway cough sensors, and the mechanical stretch and traction of fibrotic lung tissue. Cough is now being included as an endpoint in ILD clinical trials and a P2X3 antagonist has shown initial positive results in treating cough. Non-pharmacologic strategies have expanded including the use of specific lozenges and herbal mixtures, identification and avoidance of triggers, and speech and cough-suppression physiotherapy.

- Patients who are newly diagnosed with ILD, especially IPF, have often experienced a delay in diagnosis and are already experiencing significant symptoms. In addition to medical therapies, providing the patient and their family with immediate written and online resources aimed at reducing symptoms, improving mobility and socialization, and providing accurate information, gives the patient and their family critical support even when their disease outcome may not change.

Treating the dyspnea, cough, and fatigue that are commonly experienced by patients with Interstitial Lung Disease (ILD) is challenging. A multi-faceted symptom management plan offers these patients critical strategies to enhance their quality of life (QOL), especially in the setting where available medical therapies fail to meaningfully impact disease progression.

Dyspnea is a common symptom of ILD [1, 2] as well as a strong predictor of QOL in IPF patients [3-5]. The mechanisms of dyspnea in ILD include lung stiffness from scarring and inflammation, diffusion defect with hypoxemia, mismatching of ventilation and perfusion, physical deconditioning, and anxiety [2]. Regular assessment at clinic visits of resting and exercise dyspnea, as well as oxygen saturations, identifies patients with progressive symptoms requiring adjustments of their oxygen prescription, medical treatment, and goals of care. A variety of easy-to-use dyspnea scales are available that correlate with QOL [3]. The cornerstone of dyspnea management is referral to a Pulmonary Rehabilitation (PR) program which includes physical conditioning, oxygen assessment/titration, breathing retraining, and instruction in pacing, panic control, and other dyspnea self-management techniques [6]. Post PR outcomes for ILD patients consistently document significant improvements in exercise capacity, dyspnea, depression and QOL [6-15]. ILD patient and family interviews document their need and desire to discuss ILD-specific disease education, symptom management, clinical tests, autonomy, oxygen use, medications, and end-of-life counseling during PR [16, 17]. PR
provides a critical ILD treatment component that can’t be provided by an ILD patient clinic visit.

Cough can be a more troublesome symptom than dyspnea for many ILD patients. Cough occurs in more than 80% of IPF patients and also predicts a worse survival [18]. It is socially intrusive, physically exhausting, and causes distressing symptoms including dizziness, incontinence, and anxiety/panic. Patient-reported cough triggers include talking, laughing, or movement confirming the negative impact of cough on socialization and mobility [19]. Two recent reviews of cough in IPF [20] and ILD [21] include a summary of the mechanisms of cough citing increased cough reflex sensitivity or ‘up-regulation’, stimulation of nerve fibers from the fibrotic mechanical distortion of lung tissue, increased levels of sputum neurotrophins which increase capsaicin sensitivity and the cough reflex, and mechanical factors from the fibrotic lung tissue. The mechanisms for cough in IPF may overlap with those in patients with Hypersensitivity Pneumonitis and Connective Tissue Disease [21]. Causative co-morbidities of cough include gastroesophageal reflux, chronic sinusitis, obstructive sleep apnea, and use of angiotensin converting enzyme inhibitors. In a cohort of 65 IPF patients, 87% had documented acid reflux with many patients having ‘silent’ symptoms, as well as persistent reflux despite being on therapy [22].

FDA-approved and investigative cough treatments include benzonatate, baclofen, pregabalin, gabapentin, amitriptyline, nebulized lidocaine, interferon alpha, and thalidomide. While some individuals may benefit, most of these agents have not succeeded either due to lack of efficacy, side effect limitations, or small sample size [21]. Prednisone and opiates are beneficial and appropriate for select patients, especially those with advanced fibrotic disease. The successful treatment of cough in ILD patients remains elusive, but recent trials testing a P2X3 antagonist [23], cough-targeted physiotherapy [24], pirfenidone [25], and now the inclusion of cough severity as an endpoint in ILD clinical trials, all provide improved hope of added therapies. Understanding the mechanism of their cough is helpful for ILD patients as is providing them with self-management techniques such as anticipating triggers of their cough, using a lozenge before talking on the phone, avoiding irritants, increasing their oxygen flow during coughing ‘bouts’, and learning cough suppression physiotherapy techniques including vocal hygiene, speech therapy, and psycho-educational counseling [26, 27].

Effective ILD symptom management begins with early evaluation at a center with ILD expertise such as the Pulmonary Fibrosis Foundation’s (PFF) Care Center Network sites, [28, 29] followed by referral to a Pulmonary Rehabilitation program. Recent emphasis on tailoring PR to the needs of ILD patients may further improve its benefit [16, 30, 31]. Connecting our ILD patients to ILD and Caregiver support groups (in person or online), as well as patient advocacy organizations, such as

**ILD New Patient Packet:**
- ILD team contact info
- PFF patient education materials
- Oxygen education and oximeter info
- “Cough Tips” information sheet
- List of PR programs
- Support Group information
- Clinical Trial information
- Social Worker contact info
the PFF, enables patients and their families to better cope with the depression and anxiety, social isolation, and lack of accurate information. A packet of resource materials should be provided to and reviewed with all new ILD patients at their initial clinic visit. There is much to offer these patients despite many having a poor prognosis; support and education are key to optimizing their symptom management.

References:


