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Physical Therapy Management and Considerations of a Patient with Interstitial Lung Disease in the ICU: A Case Report

Timothy Mack
University of Iowa

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Timothy Mack

DPT Class of 2017
Department of Physical Therapy & Rehabilitation Science
The University of Iowa

Abstract

Background: Interstitial lung diseases (ILD) refer to a group of diffuse parenchymal lung diseases classified together based on common radiographic, physiologic, or pathologic manifestations. Although there are many different classifications, the hallmark of ILD is fibrosis of alveolar walls, airways, and vasculature resulting in a restrictive lung disease. Some research supports the benefits of pulmonary rehabilitation for sub-acute treatment of ILD, but there is even less information regarding physical therapy interventions for acute exacerbations. **Purpose:** The purpose of this case report is to present interventions and outcomes for a patient with ILD in an intensive care (ICU) setting, as well as other considerations and interventions to consider with this patient population. **Case Description:** A 64 year-old female was admitted to the ICU with hypoxic respiratory failure. The patient's past medical history was significant for ILD, systolic heart failure, severe pulmonary hypertension, coronary artery disease, oxygen dependence, obstructive sleep apnea, and hypothyroidism. Physical therapy interventions focused on increasing walking distance, with close monitoring of the patients vitals and respiratory status to ensure safety. **Outcome measures:** The primary outcome measure used during the patient's hospital stay was walking distance, showing an improvement from 15 feet to 200 feet over 20 days. **Discussion:** This case presents an acute plan of care for a patient with ILD in the ICU setting, addressing efficacy of exercise training in this patient population, and demonstrating success in improving walking distance in this patient case in preparation for discharge to sub-acute pulmonary rehabilitation.

Background

Interstitial lung diseases (ILDs) are a heterogeneous group of diseases that affect the parenchyma of the lung. The parenchyma includes the alveoli, alveolar epithelium, the capillary endothelium, and space between these structures. As a group, they are classified together based on similar clinical, pathophysiologic, and medical imaging manifestations. The term “interstitial” can be somewhat of a misnomer, as it refers to a common finding of interstitial opacities shown on a chest radiograph and the pathologic appearance that the disease first affects the interstitium. [1] However, most of the over 200 diseases classified as ILDs, have significant involvement of alveoli and airway architecture. ILDs are classified into two groups, ILDs of known cause and idiopathic ILDs. The most common known causes of ILDs are inhalation of occupational and environmental agents, most notably inorganic or organic dusts, pulmonary toxicity due to drug reaction, and radiation-induced lung injury. Various connective tissue diseases can also lead to ILD such as polymyositis/dermatomyositis, rheumatoid arthritis, systemic lupus erythematosus, and scleroderma. Accurate diagnosis is important because medical treatment of the disease will vary based on different types of the disease. Diagnosis usually requires a team of healthcare professionals working together. Regardless of classification, the hallmark of ILDs is irreversible fibrosis of the lung parenchyma. [2]

Precise pathogenesis of ILD is poorly understood. A popular proposal involves damage to distal airspace tissue from environmental exposure, systemic disease, infection, or radiation. [3] Another similar proposal, seen in **Figure 1**, involves distal airspace tissue damage but suggests the body responds to this damage with abnormal wound healing, leading to fibrosis. [2] Fibrosis leads to many detrimental factors for these patients and can progress to a vicious cycle. Oxygen diffusion limitations and ventilation-perfusion mismatching occur as the pulmonary capillary bed is damaged. This leads to patients experiencing dyspnea on exertion, which can lead to decreased activity levels. Decreased activity levels lead to further physical deconditioning and more pronounced dyspnea on exertion. Pulmonary vasoconstriction may also result in pulmonary hypertension and possible cardiac dysfunction. [4]

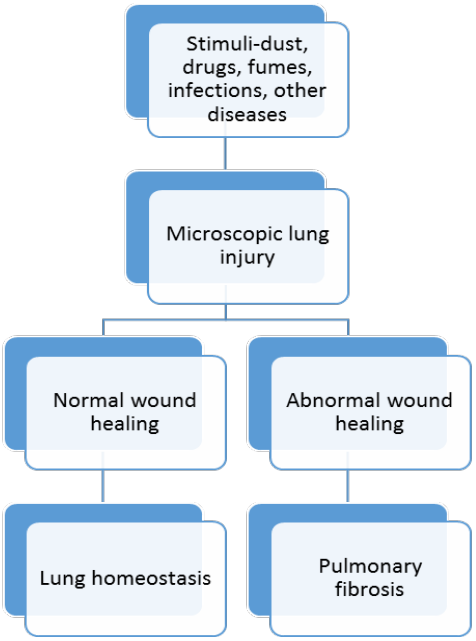

Figure 1. Pathogenesis of pulmonary fibrosis. Adapted from D.L. Kasper, A.S. Fauci, S.L. Hauser, D. L. Longo, L. Jameson.

Medical treatments for ILD are limited, as the disease is progressive with no known cure. The main goal is typically to remove offending agents or inflammation that lead to lung tissue damage. High doses of corticosteroids and antibiotics are commonly prescribed even though there is little evidence to show efficacy of these treatments. [2] Long term use of corticosteroids may have negative impacts on muscle function [5]. Pulmonary fibrosis is largely irreversible so limiting the extent of damage is key for good prognosis. [4] However, recent studies have shown potential benefits of medications such as pirfenidone and nintedanib. These benefits include slower decline of forced vital capacity (FVC) and reduced mortality. [6] Supplemental oxygen is often prescribed for patients with advanced ILD who have resting hypoxemia, but lack of evidence has made it difficult to know if supplemental oxygen has resulted in clinically meaningful outcomes [7]. Ultimately, lung transplantation is often considered for patients with severe progression of the disease. [2]

Patients with ILD are prone to acute exacerbations which are associated significant morbidity and mortality. Patients often present to the clinic with severe hypoxemia and respiratory failure and require admission to the intensive care unit. [8] Hospitalization is important for medical management but may lead to further deconditioning. One study found during an acute hospitalization, that older adults spend approximately 83% of their stay in bed and 12% in a chair. This increase in inactivity can lead to declines in strength, muscle mass, and physical function [9]. Patients with ILD may be more susceptible to these declines as their endurance and exercise tolerance is already limited. Minimal research is available regarding most appropriate acute physical therapy interventions in this patient population. It may be difficult to work with these patients in an acute setting because their symptoms are most likely at their worst during this time. Yet, physical therapists have an opportunity to promote physical activity in this setting, which if tolerated, may lead to less deconditioning. This will potentially lead to a smoother transition from hospital care to home and outpatient pulmonary rehabilitation. Thus, the purpose of this case is to present an acute plan of care for a patient with ILD in the ICU setting and address potential outcomes of exercise training in this patient population.

Case Description

Presentation/History

A 65 year old female presented to pulmonary rehabilitation for a six minute walk test (6MWT) via motorized scooter. She reported severe shortness of breath (SOB) after using the bathroom, and transferring from the scooter to the toilet and back. Vitals were obtained revealing oxygen saturation (SpO2) of 69% for both forehead and finger probes, heart rate was 127 beats per minute, and systolic blood pressure was 82 mmHg via Doppler, as cuff blood pressure was difficult to hear. The patient was normally on 5 liters per minute (LPM) of supplemental oxygen at baseline. This was increased to 6 LPM via high flow nasal cannula and then to 8 LPM to improve SpO2 to the low 90's at rest and mid 80's when the patient talked. A pulmonologist was notified and the patient was agreeable to be admitted to the ICU for further monitoring of acute hypoxic respiratory failure.

In the ICU, the patient required 10 LPM of oxygen at rest to maintain adequate Oxygen saturation, but continued to desaturate with minimal activity such transferring to the bedside chair and talking. Computed tomography (CT) of the chest showed slight progression of ILD when compared to a chest CT from two years prior. A ventilation and perfusion (V/Q) scan showed a positive mismatch, indicating possible pulmonary embolism (PE). Computed tomography angiography (CTA) ruled out a PE, so V/Q mismatching was postulated to be secondary to pulmonary fibrosis. An echocardiogram revealed right to left atrial shunting and right ventricular enlargement. A right heart catheterization was performed and showed significant pulmonary hypertension. Sildenafil and diuretics were started to help manage her pulmonary hypertension. Ambrisentan was started two days after sildenafil for optimum pulmonary hypertension management.

The patient’s past medical history was significant for ILD of unknown cause, oxygen dependence, systolic heart failure, morbid obesity, severe pulmonary hypertension, obstructive sleep apnea, coronary artery disease, and cerebral vascular accident (CVA) in 1997.

Previous pulmonary function tests (PFT’s) collected 5 months prior to hospitalization are provided in **Table 1**. Her PFT’s reveal typical findings for a patient with ILD. [2]

The patient reported living with her husband in a multi-story home. She was able to stay on the main floor and didn’t have to go up steps to get into her home. Her pre-admission level of function was walking very short distances in her home only. She reported no use of assistive device for ambulation and utilized a motorized scooter when going outside the home. The patient reported being able to walk very short distances before becoming short of breath (SOB).

Table 1. Pulmonary function test data collected 5 months prior to hospitalization.

	Measured Value (liters)	Predicted value (liters)	Percent of predicted value
FVC	.96	1.99	48
FEV1	.96	1.54	62
TLC	Not measured	3.87	

(FVC) forced vital capacity, (FEV1) forced expiratory volume in 1 minuet, (TLC) total lung capacity

Evaluation

A physical therapy evaluation was performed on the second day of the patient's hospital admission. Her vitals were stable on 10 LPM of oxygen with SpO2 reading at 96%. Her gross range of motion was within functional limits bilaterally. The patient demonstrated mild weakness with upper extremity and lower extremity strength testing. Diminished breath sounds were heard with auscultation of the lungs. Lung examination is often abnormal in patients with ILD but findings vary and are usually nonspecific. Crackles are commonly heard and may be present only in lung bases in early stages of the disease. It may be helpful to auscultate the lung bases in the posterior axillary line [1]. Breathing rate was increased with shallow breaths consistent with reports from the literature indicating patients with pulmonary fibrosis exhibit rapid and shallow breathing with low values of tidal volume and very high breathing frequencies. [10] The patient was independent with bed mobility and only required standby assistance with transfers and ambulation. Assessing the patient's gait revealed slow gait speed but no significant gait deviations. The patient demonstrated significant cardiovascular deconditioning. She was able to walk a distance of 10 feet before requiring a seated rest break due to SOB rated at 5/10 and fatigue. Careful planning was required to manage ICU equipment and supplemental oxygen to ensure the safety of our patient. "6-Clicks" basic mobility screening was calculated to be 19. This mobility screen is a short form adapted from researchers at Boston University. The form reports how much assistance the patient needs for six basic mobility questions. Higher scores indicate higher mobility with minimum score of 6 and a max score of 24. [11]

Clinical Impression

The patient would benefit from physical therapy to improve walking distance and conditioning with safe monitoring of vitals and oxygen requirements. The goal was to maintain aerobic endurance in order to return home and attend outpatient pulmonary rehabilitation.

Interventions

The patient was seen by multiple physical therapists at a frequency of 4-6 times per week over her three week period of hospitalization. Interventions focused on keeping the patient as active as possible to minimize physical deconditioning while hospitalized.

Several treatments were used with input based on clinical judgement of a senior therapist with extensive experience working with pulmonary patients. However, ambulation was our main intervention. Initially, the patient was very limited in her ability to ambulate even short distances. Her SpO2 would drop to the mid 80's and she would complain of SOB. After the initial evaluation, a front-wheeled walker was used to promote conservation of energy. Progress with walking distance was slow and hindered by continuous oxygen desaturations and SOB with exertion. We also incorporated various treatment strategies throughout the plan of care. We implemented interval walking to avoid extensive desaturation and feeling of SOB.

The patient was unwilling to ambulate at times during her first week of hospitalization because she felt tired and was scared of making her symptoms worse. When this occurred, she performed

Theraband ® resistance exercises in bed, targeting both upper extremity and lower extremity muscles groups.

The patient was educated on breathing strategies to optimize her available lung function. We recommended avoiding slow, deep breathes during ambulation because fibrotic lungs exhibit decreased compliance and will require more force to expand, resulting in increased work of breathing. Increased work of breathing results in respiratory muscles requiring a larger portion of total metabolic energy available. It is possible this leads to less total energy available to fuel muscles associated with ambulation. [10] Instead, we recommended breathing a comfortable pace, avoiding deep breaths, to halt significant energy expenditure of respiratory muscles. This may seem counterintuitive because increased respiratory rate leads to increased dead space ventilation. However, we postulated conserving energy during ambulation was more important. Working on deep breathing and expansion of lungs at rest may be more appropriate for this patient population. We also educated our patient to be cognizant of when her symptoms begin and to monitor her breathing frequency as a possible indicator of desaturation events.

Nine days after admission, the patient was medically stable and transferred to a step down unit. We continued working on progressing with walking distance until discharge.

Outcomes

Over the course of the patient's three week hospitalization, including time in both the ICU and step down unit, the patient showed improvements in walking distance, required less supplemental oxygen at rest, and demonstrated an improved "6-Clicks" basic mobility score. Measured walking distance over the patient's hospitalization can be seen in **Table 2.** At discharge, the patient was able to walk 200 ft, 190 feet further compared to admission, using a wheeled walker with several seated rest breaks but still required 15 LPM of oxygen to keep SpO2 above 90%. However, she was able to walk 30 feet on 15 LPM before desaturation began. This is an improvement of 25 ft but with greater oxygen. Resting oxygen requirements had decreased to 8 LPM. Unfortunately, oxygen requirements at rest did not return to the patient's baseline of 5 LPM. The patient's "6-clicks" mobility score improved from 19 to 22, short of the minimal detectable change of 4.72. [12]

The patient was discharged home with in home physical therapy to continue her exercise progression. There was also a recommendation made to start outpatient pulmonary rehabilitation when able.

Table 2. Progression of walking distance over course of hospitalization

Day	Walking distance (ft)
1 (admission)	10
4	50
7	100
18 (discharge)	200

Discussion

Interstitial lung diseases are a group of disabling chronic lung conditions associated with interstitial inflammation and fibrosis of alveolar walls, airways, and vasculature resulting in a restrictive lung disease. [1,13] Patients often have reduced exercise capacity and SOB during exercise which can lead to decreased quality of life. [14]

Acute exacerbations of the disease can occur at any time and often lead to hospitalization, as seen with the patient presented in this case. Frequency of exacerbations is unknown as reported incidence rates vary. Clinical presentation consists of rapid worsening of respiratory symptoms with increased dyspnea. [3] A significant secondary complication associated with ILD is pulmonary

hypertension. Pulmonary hypertension is prevalent in 30-40% of patients with ILD and is associated with high morbidity and mortality. [15]

Medical treatments are limited and often demonstrate minimal impact on quality of life. Recent studies have led to conditional recommendations for using nintedanib and pirfenidone. Pooled analysis of three randomized control trials (RCT) showed fewer number of patients with more than 10% decline in FVC over a 12 month period when taking nintedanib compared to placebo (moderate confidence). However, concerns in regard to the high cost of this treatment may limit its availability. It's also not reported whether this minimal decrease in progression of FVC decline would result in any clinically meaningful difference for these patients. Pirfendione is an oral antifibrotic drug shown to regulate important profibrotic and proinflammatory cytokine cascades *in vitro* while reducing fibroblast proliferation and collagen synthesis in animal models of lung fibrosis. Pooled results from 2 trials suggested reduced mortality (moderate confidence) and reduced rate of FVC decline (high confidence). [6]

Pulmonary function testing is one tool to help assess the progression of pulmonary damage associated with ILD. As fibrosis becomes more widespread lung volumes and compliance decrease. This restrictive manifestation results in decreased lung volumes, which in turn leads to reduced forced expiratory volume in one minute (FEV1) and forced vital capacity (FVC). The FEV1/FVC ratio often is unaffected because there is no obstruction preventing air from being expired. Destruction of the alveolar capillary bed and reduced compliance of the lung may result in V/Q mismatching, as seen in the patient presented in this case. [2]

Pulmonary rehabilitation for patients with obstructive lung diseases is well-established in the literature as improving exercise tolerance and reducing symptoms. Less evidence exists demonstrating similar effects for patients with ILD. Greater prevalence of exercise-induced hypoxia, pulmonary hypertension, and arrhythmia in this population compared to other chronic lung diseases warrants need for safe monitoring. Recent studies have shown exercise training to be safe for patients with ILD and demonstrate improvements in exercise tolerance, dyspnea, 6 minute walk distance, and quality of life after outpatient pulmonary rehabilitation. However, long-term benefits of pulmonary rehabilitation have yet to be demonstrated in the literature. This may be due to the progressive nature of the disease. [14]

There is a lack of evidence for acute physical therapy management of patients with ILD in the literature. Managing these patients in the ICU may present certain challenges not seen in other settings. Their baseline exercise capacity is often reduced and admittance to the ICU usually means they have other acute problems, likely leading to further reduced exercise capacity. Both the patient and other healthcare professionals, not familiar with physical therapists working with these patients, may have fear about making symptoms worse with physical activity. Managing that fear may consist of patient education and presenting the patient with alternative forms of exercise. Managing ICU equipment while working with critically ill patients can feel overwhelming, especially for a physical therapist with minimal experience in this setting. Planning ahead is crucial to ensure safety for the patient. The physical therapist must manage patient lines and supplemental oxygen, monitor the patient's vitals, and have options for the patient to sit when a seated rest break is needed. Assistance from nursing or other medical professionals may be required in some instances to ensure safety for these patients.

The patient in this case showed gradual improvement in her walking distance over the course of her hospitalization. We emphasized keeping the patient as mobile as possible, while safely monitoring her symptoms to avoid further exacerbation of her symptoms. She was able to tolerate some form of treatment on most days, which lead to her gradual improvement overall and eventual discharge home. She was also given a recommendation to attend outpatient pulmonary rehabilitation. This case supports the value of physical therapy in the ICU setting to promote safe exercise progression for patients with ILD.

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