Keeping Patients Active

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Alpha, Antitrypsin Deficiency (AAT) is a genetic condition, passed from parents to their children. This inherited disorder increases the risk of developing Chronic Obstructive Pulmonary Disease (COPD), liver disease, and several other conditions. The deficiency of the protein, alpha, antitrypsin, can lead to emphysema and is often misdiagnosed as asthma or smoking-related COPD. AAT deficiency related lung disease presents with common respiratory symptoms including shortness of breath, chronic cough and sputum production, wheezing, decreased exercise tolerance, frequent lower respiratory tract infections, bronchiectasis and history of suspected allergies and/or asthma. Although previously considered a disease of Caucasians, recent data show that AAT deficiency exists in all racial subgroups worldwide. While there is no cure for AAT deficiency, several effective treatments are available including smoking cessation, management of COPD/emphysema and other complications, and augmentation therapy with purified alpha, antitrypsin protein.

Exercise and activity limitation are characteristics features of COPD. This intolerance to exercise results in ventilator limitations, cardiovascular impairment and/or skeletal muscle dysfunction which can ultimately lead to a sedentary lifestyle, limited physical function, and diminished quality of life.

Various types of exercise training involving both lower and upper extremities, and respiratory muscles have been shown to improve muscle function, exercise endurance, and quality of life, while decreasing the level of dyspnea for patients with COPD. Following a regular, sustained exercise program should be part of a comprehensive therapy for lung disease patients as this has shown to increase 6-minute walk distance, exercise capacity, and several indicators of quality of life.

A recent study was conducted at the Los Angeles Biomedical Research Institute at Harbor-UCLA Medical Center (LA BioMed) using the Breathe Technologies Non-Invasive OPEN Ventilation (NIOV) System. The NIOV System is an ultra-light weight, wearable, volume augmentation ventilation system, designed to help improve breathing and facilitate mobility.

This study evaluated the physiologic effects of using the NIOV System during constant work rate exercise in subjects with COPD. Providing both supplemental oxygen and augmented ventilation, the NIOV System has the potential to promote rehabilitation and improve the health-related quality of life (QOL) in patients with respiratory insufficiency. From previous studies on the NIOV System, subjects have reported that while using the system they experienced less dyspnea, reduced work of breathing and greater mobility and exercise endurance compared to their current oxygen only systems. Physical activity is an important clinical parameter related to morbidity and mortality and is potentially a key aspect to target as an outcome measure for the NIOV System. Improving physical activity allows the patient to better participate in daily life and perhaps more importantly may spin-off long term-health benefits.

Dana Jones, an AAT deficient patient, was initially introduced to the NIOV System when he volunteered to participate in the LA BioMed study. Mr Jones experienced such a dramatic improvement in his shortness of breath and exercise tolerance while using the NIOV System, that he immediately requested a prescription to enable him to use the NIOV System at home.

Mr Jones was diagnosed with AAT deficiency in 1999. Within a year after diagnosis, he initiated AAT augmentation therapy with weekly intravenous Prolastin infusion. As his disease and symptoms progressed, by mid-2009, Mr Jones required supplemental oxygen and was experiencing respiratory insufficiency even during mild physical activity. Using the NIOV System, his shortness of breath and fatigue were noticeably reduced. Mr Jones understands that exercise is essential and that lack of it can lead to both central and peripheral deconditioning, and as a result he could experience greater dyspnea and intolerance to exertion, and further loss of functional capacity. Thus, to improve his mental outlook, stamina, and physical wellbeing, Mr Jones routinely exercises.

As dyspnea and exercise capacity worsen, the need for medical care increases and the patient’s ability for self-care decreases. These functional limitations and dependence on others can lead to a sense of loss of control, with consequent depression and anxiety. In Mr Jones’ case, the portable NIOV System provides utility in improving mobility and exercise tolerance. By optimizing conditioning, and supporting ventilatory function at various levels of physical activity, the NIOV System has the potential to improve health-related QOL, while promoting participation in activities of daily living.

Mr Jones now routinely uses the NIOV System during exercise sessions with his respiratory therapy group. Endurance exercise on the treadmill in conjunction with NIOV therapy has helped improve Mr Jones’ aerobic capacity, ventilatory muscle function, and his skeletal muscle performance. The NIOV System allows patients to select from three volume settings and can therefore be used during...
both high and low-intensity exercise. When comparing his exercise endurance using a traditional nasal oxygen cannula to the NIOV System, Mr. Jones has noticed a dramatic improvement in his exercise endurance.

Resistance exercise, like lifting weights, has the potential to improve thoracic cage muscles of ventilation and the ability to perform the activities of daily living at home, work, and play. The picture gallery includes a photo showing Mr. Jones using the NIOV System to match his ventilatory and oxygen needs during weight lifting. Using the NIOV System during resistance exercise has decreased his symptoms of dyspnea and fatigue.

Patients with chronic lung disease frequently experience activity restrictions and discomfort during activities of daily living (ADLs). For most individuals with moderate-to-severe chronic lung disease, even basic daily activities can be strenuous and daunting. For Mr. Jones, ADLs including walking stairs, carrying and lifting objects, bending down and rising from a seated position have been significantly enhanced using the NIOV System. For many patients, including Mr. Jones, bathing, dressing, and grooming require a great deal of energy consumption and oxygen use. While performing these daily tasks, especially showering and shaving, Mr. Jones reports experiencing significantly less dyspnea and greater control of his breathing with the NIOV System.

In other photos, Mr. Jones can be seen working on his classic 1971 Mini Cooper, which he cherishes greatly. The NIOV System has enabled him to continue his hobbies and given him greater independence to perform the activities he enjoys most.

Chronic lung diseases, like ATT deficiency, are progressive diseases that slowly rob patients of exercise tolerance, health-related QOL, and independence. Exercise in conjunction with education and other components of formal pulmonary rehabilitation have clearly been shown to improve functional status, dyspnea, and health-related QOL in patients with lung disease. A higher level of physical activity can reduce the number of hospital admissions due to exacerbations, as well as decrease respiratory mortality. For Mr. Jones, the NIOV System has significantly increased his functional exercise level of daily physical activities by reducing his work of breathings while effectively supplementing his oxygen intake. The system has not only improved Mr. Jones’ independence and security, it has also made a significant improvement in his level of dyspnea, and exercise endurance.

References
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