Disability in Pulmonary Hypertension

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Disability in patients with pulmonary hypertension (PH) is rooted not only in physical impairment, but can also be the result of associated mental and emotional dysfunction often experienced by PH patients. Very little has been done to document the extent and degree of disability in PH, and even less has been done to determine the effect of PH treatments on disability. While current US policies governing eligibility for disability do not take into account many of the factors that lead to disability that are unique to PH patients, work is underway to revise these policies.

QUALITY OF LIFE IN PH

Although PH cannot be cured, medical therapy often allows the afflicted patient to live with the chronic illness, albeit with varying degrees of impairment. This impairment is usually physical, with dyspnea, fatigue, and exercise intolerance as the most common symptoms in pulmonary arterial hypertension (PAH) patients. However, mental impairment, emotional dysfunction, and impaired sleep quality may also contribute significantly to the impairment of PH patients. Few studies have reported quality of life (QoL) or change in QoL with treatment in PH patients. In a 2009 review of the literature on QoL in PAH, Rubenfire et al found that most PAH drug studies used generic instruments to study QoL, and point out that additional factors besides cardiopulmonary dysfunction contribute to reduced QoL, such as muscle weakness and lack of flexibility in scleroderma-related disorders. They also emphasized that physical symptoms of fatigue, weakness, and shortness of breath can be related to nonphysical factors such as anxiety and depression. In one study of 46 PAH patients, more than half had impaired cognitive function.

Depression is common in patients with PAH. Up to 50% or more of patients with PAH are depressed, with 1 in 7 having major depressive symptoms. Perhaps the first report of depression in patients with PH was in a cohort of 164 patient members of the German PH association, pulmonale hypertonie e.v. In this extensive report, 35% of patients overall had mental disorders, with the most common being major depressive disorder and panic disorder. However 62% of PH patients with NYHA Class IV symptoms suffered from mental disorders, effectively linking the degree of mental impairment with the severity of the underlying PH. Indeed, depression is associated with decreased exercise capacity in many cardiac and pulmonary disorders, and can alone lead to inability to work. Thus, both physical and mental impairment can lead to disability, and the two may feed off of each other.

PHYSICAL IMPAIRMENT IN PH

One of the hallmarks of the physical impairment due to PH is reduced exercise capacity, with exertional breathlessness (ie, dyspnea) being the most common symptom reported. Both the inability to increase cardiac output during exercise and the impairment in the ability of pulmonary blood flow to perfuse ventilated lung (resulting in poor ventilation/perfusion matching) contribute to the impaired exercise capacity, dyspnea, and fatigue experienced by patients with PH. Interestingly, however, formal measures of exercise capacity, such as with cycle ergometry, do not correlate well with functional status and activities of daily living. This may in part be related to the interplay between affected physical and nonphysical systems in PH.

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Additional side effects associated with some PH treatments include pain at the medication injection site (which in itself can be debilitating), catheter infection and sepsis requiring hospitalization and catheter removal, and coughing after inhalation of PH medications. Replacement of central venous catheters is not without significant risk in part due to the limited...
WORK DISABILITY DUE TO PH

The physical impairments, reduced QoL, and associated depression commonly experienced in PH patients contribute to a high likelihood of PH patients becoming disabled. Because PAH often affects young individuals that are part of the workforce, the inability of PAH patients to remain in the workforce is of particular concern. Unfortunately, the effect and magnitude of work disability has not been reported for these patients.

In the US, individuals with conditions that render them unfit for employment must meet eligibility requirements for financial benefits, set forth by the policies of the Social Security Administration (SSA). The SSA considers an individual disabled if he/she is unable to work in the job that he/she did prior to becoming disabled, and cannot adjust to other types of work because of the medical condition(s). The disability must also be expected to last for at least one year or to result in death.

The current eligibility criteria for PH patients to be certified as disabled are addressed in the SSA’s Blue Book in several areas, including Section 3.00: Respiratory System, subsection 3.09: “Cor pulmonale secondary to chronic pulmonary vascular hypertension,” and is defined as follows:

1. Mean pulmonary artery pressure greater than 40 mm Hg;
2. Arterial hypoxemia. Evaluate under the criteria in 3.02C2.

WORK DISABILITY AND RESPONSE TO TREATMENT

None of the pivotal studies involving pharmacologic PAH treatments have reported the prevalence of work disability to date, and thus none have reported on the successes or failures of these treatments in reducing work disability. Surgical intervention for chronic thromboembolic pulmonary hypertension (CTEPH), however, would seem to be another highly successful treatment for PH patients.
has been shown to facilitate return to work. Archibald et al.\textsuperscript{16} reported that 43% of patients undergoing pulmonary thromboendarterectomy (PTEA) returned to work an average of 16 weeks after surgery; 62% of these patients had not been working prior to surgery.

While PTEA may be more effective at alleviating the symptoms of PH than pharmacologic PAH treatments, it is not unreasonable to hypothesize that some PAH patients might respond sufficiently to treatment such that they are enabled to return to work. As PAH treatment further evolves, analysis of disability-related outcomes will likely prove useful.

**CONCLUSION**

Disability is common in PH patients, and is related to several factors underlying their disease process, including physical and cognitive impairment, as well as impaired QoL. Objective statistics quantitating the nature and degree of disability in PH patients are lacking, and criteria for determination of disability fall short of accurately capturing the elements needed for disability determination. Patients, medical professionals, and the PH community at large should continue to advocate strongly for system-wide improvements in the disability determination process, while continuing to pursue methods aimed at reducing the influence of PH on physical and mental function. Finally, formal study of disability in PH should be undertaken to better define the nature and effect of PH on disability, and to have a better understanding of what needs to be done to improve the lives of PH patients.

**References**