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.[1] However, mental impairment, emotional dysfunction,[2] and impaired sleep quality[3] 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[4] 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.[5] In one study of 46 PAH patients, more than half had impaired cognitive function.[6]

Depression is common in patients with PAH.[2] 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.[7] 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,[8,9] with exertional breathlessness (ie, dyspnea) being the most common symptom reported.[1] 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.[10] This may in part be related to the interplay between affected physical and nonphysical systems in PH.

Physical impairment in PH can also occur as a result of PAH drugs and their delivery systems (Table 1). Common side effects of PH treatments include, but are not limited to: increased thirst and urination, dry mouth, hypokalemia with muscle cramping, flushing, hypotension (ie, low blood pressure), dizziness, headache, jaw pain, leg and back pain, diarrhea, and rash.

Depending on the medications, the burden of PH treatments can include the daily need for preparing PH medications under sterile conditions; carrying ice packs to keep medication in the infusion pump refrigerated; carrying and maintaining 2 infusion pumps (one pump is needed for backup due to the short half-life of the medication, which can be as short as 3-6 minutes, thereby necessitating the 24/7 treatment); cleaning, protecting, and maintaining an indwelling central venous or subcutaneous catheter; and carrying, maintaining, and preparing a nebulizer for inhalation 4-9 times daily, with inhaled treatment delivery times ranging from 2-15 minutes per treatment. Being off the 24/7 therapies even for a few minutes can be life threatening. The burden of continuous oxygen therapy includes the need for transporting a continuous supply of oxygen (ie, not infrequently large and heavy tanks) and use of nasal cannula.

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

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Table 1: Physical Impairments Related to PH Drugs and/or Their Delivery Systems

Drug Class/Device	Contributor to Disability
Prostacyclins	Intravenous catheter, subcutaneous catheter, nebulizer, ice packs; nausea, vomiting, headache, hypotension, flushing
Oxygen	Nasal cannula, heavy tanks
Phosphodiesterase-5 inhibitors	Epistaxis, headache, dyspepsia, flushing, insomnia, erythema, dyspnea, rhinitis
Endothelin antagonists	Peripheral edema, nasal congestion, sinusitis, flushing, palpitations, nasopharyngitis, abdominal pain, constipation, dyspnea, headache

number of veins into which the catheter can be inserted.

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[11]:

Cor pulmonale secondary to chronic pulmonary vascular hypertension.
Clinical evidence of cor pulmonale (documented according to 3.00G) with:
A. Mean pulmonary artery pressure greater than 40 mm Hg;
or
B. Arterial hypoxemia. Evaluate under the criteria in 3.02C2.

Additional listings related to PH are addressed in Section 4.00: Cardiovascular System, subsection 4.06: "Symptomatic congenital heart disease," and defined as follows[12]:

Symptomatic congenital heart disease (cyanotic or acyanotic), documented by appropriate medically acceptable imaging (see 4.00A3d) or cardiac catheterization, with one of the following:
A. Cyanosis at rest, and:
1. Hematocrit of 55% or greater; or
2. Arterial O_2 saturation of less than 90% in room air, or resting arterial PO_2 of 60 Torr or less.
OR
B. Intermittent right-to-left shunting resulting in cyanosis on exertion (eg, Eisenmenger's physiology) and with arterial PO_2 of 60 Torr or less at a workload equivalent to 5 METs or less.
OR
C. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70% of the systemic arterial systolic pressure.

Unfortunately, these disability criteria are outdated and do not capture all of the essential elements of disability in PH patients, such as dyspnea, exercise intolerance, syncope, volume overload, medication/device side effects, and cognitive impairment. In addition, standardized diagnostic PH coding procedures used today for reporting and billing purposes have not always captured the true nature of the disease process, and they do not classify PH according to the World Health Organization (WHO) PH classification system.[13] Along these lines, Link et al[14] note that the few International Classification of Diseases, Ninth Revision (ICD-9) codes currently used for PH probably do not always accurately reflect the nature of the disease process.

Little is known about the success, failure, or difficulties that PH patients encounter when applying for disability. Anecdotal experience suggests that for many, perhaps the majority, the process is not always smooth, and can often take more than a year to sort out the issues. This is likely related to the nonspecific nature of PH symptoms and the inadequate SSA definitions mentioned above. Rubenfire et al note that most PAH patients have difficulty in qualifying for medical disability.[4] They suggest reasons for this difficulty, such as the rarity of the disorder, their young age, the relatively normal physical examination, and, as mentioned above, the lack of criteria specific to diseases of the pulmonary vasculature.

Fortunately, with the assistance of the Institute of Medicine, members of the PH community, and input from the public at large, the SSA plans to update the cardiovascular disability listings in the near future.[15] In addition, the Pulmonary Hypertension Association (PHA) recently commissioned an Insurance Advocacy Committee. This committee's mission in serving PH patients is, in part, to guide PHA's response to time-sensitive PH policy changes among public payers, government institutions, and specific health care fields.

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,

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has been shown to facilitate return to work. Archibald et al[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.

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