

Understanding Disability in Parkinson's Disease

Lisa M. Shulman, MD*

Department of Neurology, University of Maryland School of Medicine, Baltimore, Maryland, USA

Abstract: Even with optimal therapy, it is inevitable that the symptoms of Parkinson's disease (PD) progress and gradually result in disability in the performance of daily activities. Delay and prevention of disability is among the highest priorities in the clinical management of PD. Understanding the association between the diverse symptoms of PD and the emerging disability is fundamental to minimizing functional limitations. This article differentiates disability

from impairment and quality of life and explores the relationship between the specific impairments of PD and resulting disability. Identifying appropriate tools for outcomes measurement and impediments to accurate assessment of disability are also reviewed. © 2010 Movement Disorder Society

Key words: Parkinson's disease; disability; function; impairment; outcomes measurement

The onset of disability in the performance of daily activities is often pivotal in the diagnosis and management of Parkinson's disease (PD). Difficulty with handwriting may trigger the initial office visit and functional status is generally the main determinant of the need for antiparkinsonian medication. It is easy for the clinician to observe the trajectory of symptoms overtime and the consequence of emerging disability. This article will "drill down further" to understand the relationship between PD symptoms and disability, and will focus on the following questions. Which activities of daily living (ADLs) are most affected in PD? What symptoms of PD have the greatest impact on daily function? How accurate are self-reports of disability?

DEFINITIONS: DISABILITY VERSUS IMPAIRMENT AND QUALITY OF LIFE

The medical literature often employs terms describing disease severity like impairments, symptoms, disability, function, and quality of life interchangeably

although their definitions are distinct. *Impairments* are the symptoms and signs of a disease process. The impairments of PD include tremor, bradykinesia, freezing of gait, depression, and fatigue. Information about impairment comes from both subjective and objective sources (history and physical examination).

The World Health Organization defines *disability* as "any restriction or lack of ability to perform an activity within the range considered normal for a human being due to an impairment."¹ Disability can also be assessed based on subjective and objective data. Inquiries about performance of basic ADLs including dressing and bathing or instrumental activities of daily living (IADLs) such as shopping and preparing food, result in the patient's perspective of their disability. Objective data about disability is often also assessed during the office visit, such as when the patient is unable to undress themselves for the examination or handle toileting independently. Objective quantitative data regarding function may be collected with physical performance measures, including timed gait testing or simulations of daily activities.

Quality of life is a complex measure that comprises elements of impairment and disability, but also encompasses the patient's subjective perception of their health and well-being. By definition, quality of life is a subjective measure; understanding the subjectivity of quality of life data is the key to understanding its

*Correspondence to: Dr. Lisa M. Shulman, Professor of Neurology, University of Maryland School of Medicine, 110 S. Pace St., Room 3-5-127, Baltimore, MD 21210.
E-mail: lshulman@som.umaryland.edu

Potential conflict of interest: Nothing to report.

Received 6 December 2007; Revised 1 October 2008; Accepted 21 August 2009

Published online in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/mds.22789

strengths and weaknesses as an outcome measure. The individual's view of their health (health-related quality of life) is influenced by numerous factors, including expectations, mood, responsibilities, and personal goals. Contrast the potential impact of a new diagnosis of PD on the quality of life of a 50-year-old construction worker versus a 70-year-old retiree. Quality of life reflects a composite of multiple factors that will differ for every individual. The idiosyncratic nature of quality of life can result in perplexing and seemingly contradictory data. Nevertheless, it is difficult to quarrel with the patient being the final arbiter of the success or failure of their health care.

Distinguishing impairment, disability and health-related quality of life can be tricky. For example, gait impairment may comprise description of postural changes, stride length, arm swing, and symmetry, whereas disability when walking will mainly focus on the capacity to comfortably walk routine distances during normal daily activities. In contrast, a person's perception of their walking (health-related quality of life) may encompass their personal values, priorities, and perspectives. For example, an alteration of posture or the presence of dyskinesia during ambulation may result in embarrassment and less quality of life even though there is no actual limitation of mobility.

DISABILITY AND PD SEVERITY: IS THERE REALLY A DIFFERENCE?

It is well recognized that disability in PD is correlated with both disease severity and quality of life. At the University of Maryland PD and Movement Disorders Center, patients visiting the center are routinely assessed for impairment with the Unified Parkinson's Disease Rating Scale (UPDRS),² disability with the Older Americans Resource and Services Scale (OARS),³ and health-related quality of life with the SF-12v2 Health Status Survey.⁴ Analysis of data from nearly 800 PD patients shows that disability strongly correlates with both disease severity ($r = 0.64$, $P < 0.001$) and quality of life in PD ($r = -0.46$ to -0.62 , $P < 0.001$). But when the relationships between disability, impairment, and quality of life are represented as individual data points instead of simple correlations, a different picture emerges (Figs. 1 and 2A,B). Although all three outcome measures are correlated with each other, there is wide variation between the level of disability that corresponds to a specific level of disease severity or quality of life. For example, in Figure 1 the UPDRS III Motor examination score

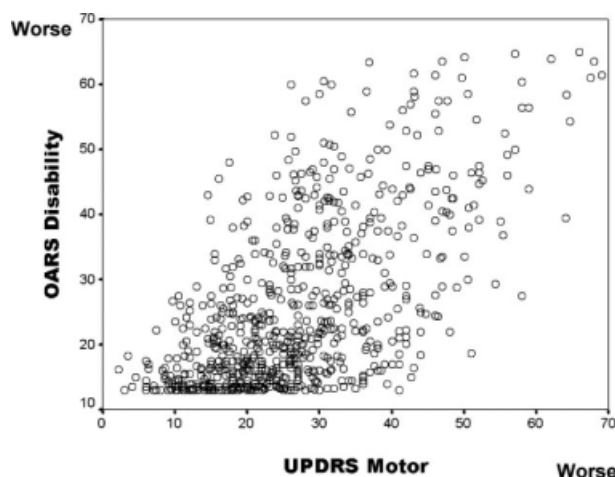


FIG. 1. A scatterplot of 759 subjects with Parkinson's disease showing the individual datapoints of self-reported disability on the OARS scale and the objective physician rating of impairment on the UPDRS III: Motor examination ($r = 0.635$, $P < 0.001$).

ranges from 5 to 40 among patients reporting no disability. Similarly, if we focus only on the patients scoring between 30 and 40 on UPDRS III, the subjective OARS disability ratings range from the lowest possible score of 14 (no disability) to over 50. This range encompasses levels of completely normal function to significant loss of independence. Even greater variability is seen in the quality of life ratings at selected levels of disability (Fig. 2A,B).

These comparisons emphasize the significance of choosing outcome measures that are targeted to specific needs. For example, a general disease severity measure such as the UPDRS is not likely to adequately reflect the benefits of a rehabilitation program or the introduction of a hypnotic for sleep disturbance. A measure of disability with sufficient sensitivity to changes in performance of ADLs and IADLs is a good match to assess the outcome of the rehabilitation program. Symptom-specific measures that focus on sleep and fatigue are good choices for assessment of the new hypnotic. The introduction of highly effective dopaminergic medication such as levodopa (L-dopa) may result in global improvements of impairments, disability, and quality of life. However, there are a number of settings where idiosyncratic results are likely. For example, the introduction of L-dopa in patients with early symptoms may improve tremor and bradykinesia; however, there may be little or no baseline disability in daily activities (hence, no improvement in disability). Conversely, L-dopa may not be effective for the substantial disability resulting from symptoms of freez-

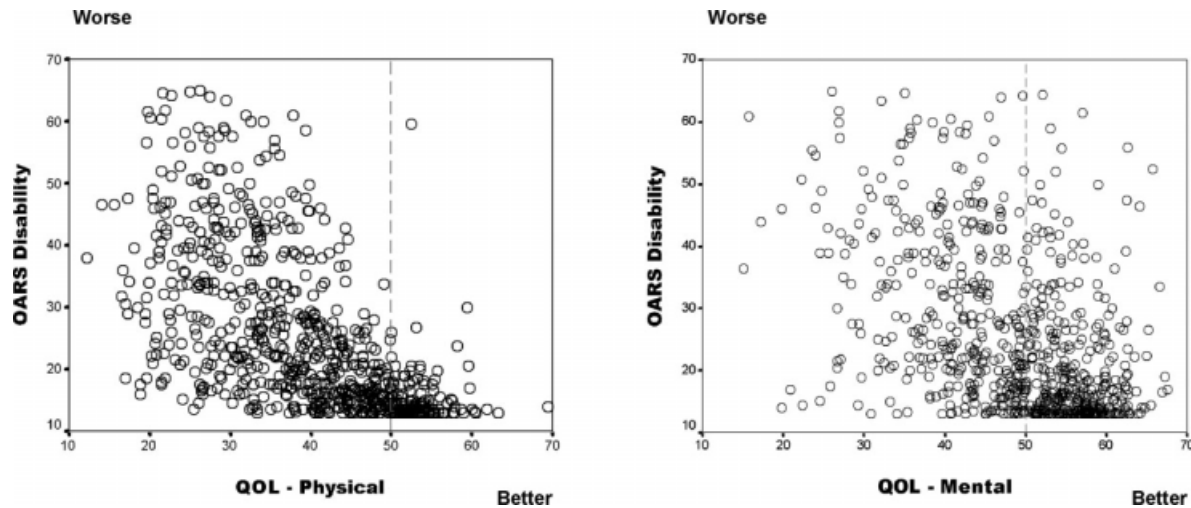


FIG. 2. A scatterplot of 744 subjects with Parkinson's disease showing the individual datapoints of self-reported disability on the OARS scale and health-related quality of life on the SF12 Health Status Survey [(A): Physical health summary score $r = -0.620$, $P = 0.001$ and (B) Mental health summary score $r = -0.457$, $P < 0.001$]. The dashed vertical line shows the 50th percentile, which is the mean score of the United States normative population.

ing of gait with loss of postural instability. In this situation, the UPDRS may markedly improve (due to reduction of tremor, bradykinesia, and rigidity), but disability and quality of life may remain relatively unchanged.

THE RELATIONSHIP BETWEEN PD SYMPTOMS AND DISABILITY

Assessment of the impact of PD on disability is included in most clinical trials, although the disability scales commonly used have limitations. The Schwab and England Activities of Daily Living⁵ scale (S&E) is a single global assessment of dependency in ADLs. The UPDRS Part II (ADL) is mostly comprised of ratings of impairments (sensory complaints, tremor, salivation) with a lesser emphasis on a subset of ADLs (walking, dressing, handwriting) and omitting the major ADL of toileting. The IADLs include shopping, meal preparation, housework, medication, and money management, but assessment of IADLs is rarely included in studies of PD, although they may be more sensitive to early disability than ADLs, and are a good indicator of dependency with the need for social services.

In addition to general disease severity, previous studies have shown that depression,⁶⁻⁹ motor fluctuations, dystonia,¹⁰ and later age of onset of PD¹¹ strongly correlate with disability. Drug-related dyskinesia had a moderate correlation and only weak correlations were observed with orthostatic hypotension and sleep disturbance.¹⁰ Recent studies of disability at the

University of Maryland PD Center show that both motor and nonmotor symptoms are important determinants.^{12,13} Among the motor symptoms, gait impairment, postural instability, and bradykinesia have the strongest correlation with disability. Tremor showed no correlation with disability, demonstrating the difference between the impact of a postural/kinetic tremor (as in essential tremor) and the resting tremor of PD. Among the nonmotor symptoms, cognitive dysfunction, psychotic ideation, urinary incontinence, motivation, and depression all had strong correlation with disability.

The study, evolution of disability in PD investigates the level of disease severity associated with disability in common ADLs and IADLs.¹⁴ A cross-sectional analysis of the relationship between levels of disease severity and disability on ADLs and IADLs was performed on 658 patients with PD. Disease severity was measured with the total UPDRS² and disability with the OARS.³ For each 10-point increment of the UPDRS, the percentage of people reporting functional limitations on the individual ADLs and IADLs were calculated. The results showed that difficulty walking was associated with the lowest, earliest UPDRS ratings (total UPDRS < 20). A number of gait-dependent ADLs and IADLs including dressing, transferring, housework, traveling, and shopping followed closely behind at UPDRS ratings between 20 and 40. Difficulty with more cognitively based IADLs including using the telephone and managing medications and money were associated with more advanced UPDRS ratings (total UPDRS 50–60).

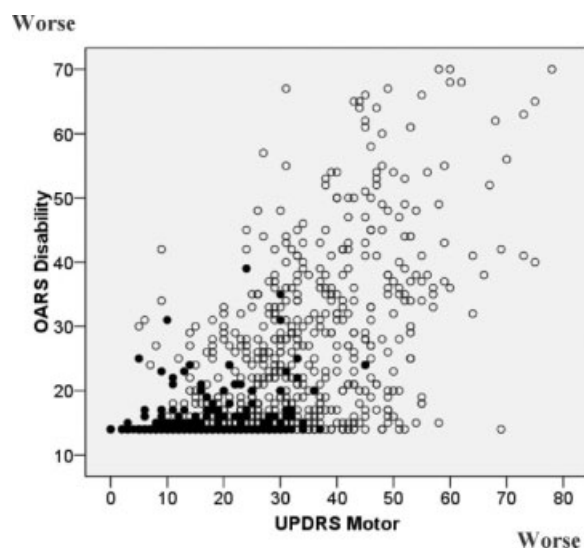


FIG. 3. A scatterplot of 965 subjects with Parkinson's disease showing the individual datapoints of self-reported disability on the OARS scale and the physician rating of impairment on UPDRS III: Motor Examination ($r = 0.611$, $P < 0.001$). The solid circles ● indicate individuals with no gait or balance impairment. The open circles ○ indicate individuals scoring greater than zero on UPDRS items 29: Gait or 30: Postural instability.

The pivotal role of gait and balance in PD-related disability is shown in the graph in Figure 3. When you code the individual datapoints of the scatterplot according to the presence or absence of gait or balance impairment, the impact of loss of ambulation on disability becomes apparent. Compare this figure with the “uncoded” identical graph in Figure 1. This illustrates how the variability in disability at levels of disease severity is explained by factors that strongly correlate with disability in PD. Gait and balance are key determinants of disability in PD.

IMPEDIMENTS TO ACCURACY OF DISABILITY ASSESSMENT

Disability assessment (as well as symptom severity) is often based on patient-reported data. The accuracy of self-reported data may be limited by many factors including cognitive function (memory, judgment, insight) and emotional state. Environmental factors also play a very important role. For example, compare two individuals with a 10-year history of PD and similar levels of gait/balance impairment. The first person lives with several supportive family members in a house on one level. The second person lives alone in a two-story home. Disability scale items such as: Are you able to dress yourself, do the housekeeping or pre-

pare the meals may result in very different responses. The first person may hardly recognize that his/her performance relies on others who do the laundry, lay the clothing out and do the food shopping. The second person may have the capacity to be more independent with the proper environmental supports.

In a recent study of subjective versus objective assessment of ADLs in PD, we found discordance between subjective and objective assessment in the majority of patients.¹⁵ Although most study subjects under-rated their objective disability, there was a tendency for those with early PD to under-rate the most, whereas those with advanced PD tended to over-rate their disability. Patients who rated themselves as more disabled than the objective observer tended to live alone, whereas patients who reported less disability than the observer lived with spouse or other family members. Other studies have highlighted the difference between patient versus “proxy” reported data.¹⁶ Although clinical practice generally involves collecting information from either patient or family members, studies consistently show that proxies (spouse and other family) tend to rate disability higher than patients.

CONCLUSION

Preventing and delaying disability is of highest priority in the management of PD. This article reviews a number of issues that are fundamental to the assessment and management of disability in PD. Recognition of limitations in the accuracy of self-reported disability should result in work to improve the reliability of data on functional status. Understanding the pivotal role of gait and balance in daily function has important clinical application. Difficulty with ambulation is a clinical “red flag” that anticipates emerging disability and should prompt evaluation for change of management. Identifying new effective interventions for symptoms that result in greatest disability is a priority. The association between the specific symptoms of PD and disability in daily activities illustrates which manifestations are likely to be most clinically meaningful to our patients.

REFERENCES

1. World Health Organization. International classification of impairments, disabilities, and handicaps. Geneva; 1980.
2. Fahn S, Elton RL, Members of the UPDRS Development Committee. Unified Parkinson's disease rating scale. In: Fahn S, Marsden CD, Calne DB, Goldstein M, editors. Recent developments in Parkinson's disease, Vol. 2. Macmillan Health Care Information; 1987. p 153–164.

3. Duke University Center for the Study of Aging and Human Development. Multidimensional functional assessment: the OARS methodology. A manual, Second ed.; 1978.
4. Ware JE, Jr, Kosinski M, Keller SD. A 12-item short-form health survey: construction of scales and preliminary tests of reliability and validity. *Med Care* 1996;34:220–233.
5. Schwab JF, England AC. Projection technique for evaluating surgery in Parkinson's disease. In: Gillingham FJ, Donaldson MC, editors. Third symposium on Parkinson's Disease. Edinburgh: Livingston; 1969. p 152–157.
6. Brown RG, MacCarthy B, Gotham AM, Der GJ, Marsden CD. Depression and disability in Parkinson's disease: a follow-up of 132 cases. *Psychol Med* 1988;18:49–55.
7. Starkstein SE, Mayberg HS, Leiguarda R, Preziosi TJ, Robinson RG. A prospective longitudinal study of depression, cognitive decline, and physical impairments in patients with Parkinson's disease. *J Neurol Neurosurg Psychiatry* 1992;55:377–382.
8. Liu CY, Wang SJ, Fuh JL, Lin CH, Yang YY, Liu HC. The correlation of depression with functional activity in Parkinson's disease. *J Neurol* 1997;244:493–498.
9. Siderowf A, Ravina B, Glick HA. Preference-based quality-of-life in patients with Parkinson's disease. *Neurology* 2002;59:103–108.
10. Rubenstein LM, Voelker MD, Chrischilles EA, Glenn DC, Wallace RB, Rodnitzky RL. The usefulness of the Functional Status Questionnaire and Medical Outcomes Study Short Form in Parkinson's disease research. *Qual Life Res* 1998;7: 279–290.
11. Diamond SG, Markham CH, Hoehn MM, McDowell FH, Muentner MD. Effect of age at onset on progression and mortality in Parkinson's disease. *Neurology* 1989;39:1187–1190.
12. Shulman LM, Anderson KE, Gruber-Baldini AL, Reich SG, Fishman PS, Weiner WJ. Disease-specific of co-morbid factors-which has the greatest impact on disability in Parkinson's disease. *Mov Disord* 2006;21 (Suppl 15):S509.
13. Shulman LM, Anderson KE, Vaughan CG, Gruber-Baldini A, Fishman PS, Reich SG, Weiner WJ. The impact of symptoms of advanced Parkinson's disease on daily function and quality of life. *Neurology* 2004;62 (Suppl 5):A335.
14. Shulman LM, Gruber-Baldini AL, Anderson KE, Vaughan CG, Reich SG, Fishman PS, Weiner WJ. The evolution of disability in Parkinson's disease. *Mov Disord* 2008;23:790–796.
15. Shulman LM, Pretzer-Aboff I, Anderson KE, et al. Subjective report versus objective measurement of activities of daily living in Parkinson's disease. *Mov Disord* 2006;21:794–799.
16. Snow LA, Cook KF, Lin PS, Morgan RO, Magaziner J. Proxies and other external raters: methodological considerations. *Health Serv Res* 2005;40(5 Pt 2):1676–1693.