Movement Disorders
CLINICAL PRACTICE

TASK FORCE REVIEW

Scales to Assess Clinical Features of Progressive Supranuclear Palsy: MDS Task Force Report

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PSP is a neurodegenerative disorder classically characterized by progressive postural instability with falls, supranuclear vertical gaze palsy, pseudobulbar palsy, levodopa-unresponsive parkinsonism, and frontal cognitive disturbances.[1,2] Given the increasing research interest in PSP, a review of published scales using similar methodology applied to other disorders[3–5] was commissioned for PSP by the International Parkinson and Movement Disorder Society (MDS).

Materials and Methods

Administrative Organization and Critique Process

The Steering Committee of the MDS Task Force on Rating Scales invited the lead author (D.A.H.) to form and chair a task force to critique existing PSP rating scales and place them in a clinical and clinimetric context.[3–5] Task force members conducted a literature search, selected the scales to be included in the review, and performed a structured assessment of the scales with regard to descriptive properties, clinimetric qualities, strengths, weaknesses, and overall impression (see Supporting Data 1). Initial scale selection was done by at least two members of the group, with additional scales added during meetings of the task force members and the Steering Committee. Data were extracted by one member and reviewed in depth by the task force and, subsequently, by the Steering Committee. Clinimetric quality was evaluated using all aspects available for each scale, including, but not limited to: face validity; content validity; reliability; internal consistency; and responsiveness to change. Results from the data analysis in the validation studies were contrasted to standard criteria, this way qualifying the goodness of the tested clinimetric attributes.[6,7]

Scales were classified as "Recommended" if they had been applied to PSP populations, if there were data on their use by several groups other than the scale developer, and if they had been studied clinimetrically and found to have adequate clinimetric properties in PSP (three criteria). A scale was classified as "Suggested" if the scale had been applied to PSP populations and had been found to have adequate clinimetric properties in PSP, or had been used by several groups in PSP (two criteria). A scale was "Listed" if the scale had been applied to a PSP population, but had been used rarely and had not been demonstrated to have adequate clinimetric data in PSP (one criterion). The final assessment was based on consensus among the task force members and the Task Force on Rating Scales for the MDS Steering Committee.

Literature Search Strategy

All scales designed to evaluate clinical features of PSP were included in the review and identified through a comprehensive PubMed search (through January 2011). Keywords searched were: (progressive supranuclear palsy) AND (rating scale OR psychiatric OR cognitive OR sleep OR quality of life). All results in PubMed from 1998 to 2011 (human only) were reviewed for potential inclusion (n = 1,652). To be included, each study had to have been conducted in patients with PSP, utilize a rating scale, and measure a feature of the disease. Published case reports and abstracts were excluded, and no attempts were made to locate unpublished studies. Searches of MedLine and Ovid were also conducted. Imaging studies were not included because they were

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beyond the scope of this task force. Additional scales discovered during the review process were added as appropriate.

Scale Review

Twenty-seven rating scales were identified that assess clinical features of PSP (Tables 1 and 2). Three of these scales were developed specifically for PSP; two of these assess the full spectrum of symptoms in PSP: the Clinical Rating Scale for Progressive Supranuclear Palsy (PSPRS)[8] and the Natural History and Neuroprotection in Parkinson Plus Syndromes–Parkinson Plus Scale (NNIPPS-PPS),[9] whereas one focuses on quality of life in PSP, the Supranuclear Quality of Life scale (PSP-QoL).[10] The other 24 scales are not specific for PSP and focus on motor, cognitive, psychiatric symptoms, or general health status of PSP patients. Three of the identified scales fulfilled criteria for Recommended for use in PSP (the PSPRS, the UPDRS,[11] and the Frontal Assessment Battery [FAB][12]), seven scales met criteria for Suggested, and these 10 scales are reviewed in detail. The 17 Listed scales are described in Supporting Data 1.

Recommended Scales

PSPRS

The PSPRS is a clinician-rated instrument to assess disability and severity of PSP.[8] Administration of the scale takes approximately 15 minutes. The PSPRS consists of 28 items scored on a 3- or 5-point Likert scale, with the total score ranging from 0 to 100. Each item is scored from either 0 to 4, with the exception of four items, which are scored from 0 to 2, with higher scores indicating more-severe disability or movement abnormality. Items are in six categories: daily activities (by history); behavior; bulbar; ocular motor; limb motor; and gait/midline. The scale includes comments and/or instructions for each item and word anchors to explain the ratings.

The scale has been applied in PSP (criterion 1) and used by groups other than the original authors (criterion 2).[13–15] Inter-rater reliability intraclass correlation coefficient (ICC) was 0.86 (95% confidence interval [CI]: 0.65–0.98).[8] Factor analysis with oblique varimax rotation was performed after administration of the scale to 162 PSP patients and the nonhistorical (excluding daily activities) items sorted into five factors.[8,16] Content validity has not been reported. Criterion validity was evaluated using progressivity of PSP, with a robust association between survival and PSPRS score (criterion 3).[8] The PSPRS is sensitive to change based on PSPRS score "milestones" over 1 year.[8] Data quality and scaling acceptability have not yet been addressed.

The PSPRS can be readily performed in the office and has been used by many investigators, displays good psychometric properties, and is free. However, many of the items require training and inter-rater reliability was studied using a videotaped version of the scale, without all items. The PSPRS fulfills criteria for Recommended scale (Table 1) for the global assessment of the disorder. In designating this rating, however, the task force recognizes that clinimetric testing has only been partial and would be enhanced by validity testing by additional groups.

UPDRS

The UPDRS provides a comprehensive assessment of disability and impairment in PD.[11] There are four subscales: mental status, behavior, and mood; activities of daily living (ADLs); motor examination; and complications. The UPDRS has been tested clinimetrically in PD.[17–19] The UPDRS motor section (UPDRS section III) has been applied in PSP (criteria 1 and 2).[20] Internal consistency in PSP was high (Cronbach's alpha = 0.9).[19] Five factors accounted for 64% of the sample variance: bradykinesia of the extremities; axial bradykinesia and gait; action tremor; rest tremor; and rigidity.[20] Correlational analyses among the factors revealed a low degree of association (r = 0.02–0.26).[20] Construct validity demonstrated that only factors assessing bradykinesia and gait were related to stage of disease.[20] Face validity was adequate, but further detailed confirmatory studies are needed (criterion 3).[19] The UPDRS section III is frequently used, but some specific features of PSP (such as

TABLE 1 Classification of recommended and suggested scales

Scale	Applied in PSP	Applied by Several Groups	Adequate Clinimetric Testing in PSP	Qualification
PSP specific				
PSPRS	X	X	X	Recommended
NNIPPS-PPS	X		X	Suggested
PSP-QoL	X		X	Suggested
Not PSP specific				
Motor				
UPDRS III	X	X	X	Recommended[a]
H & Y	X	X		Suggested[a]
Cognitive				
FAB	X	X	X	Recommended[a]
DRS	X	X		Suggested[a]
MMSE	X	X		Suggested[a]
Psychiatric				
NPI	X	X		Suggested[a]
Health status				
EQ-5D	X	X		Suggested[a]

For an explanation of the qualification groups, see text.
[a]For assessment of specific aspects of PSP.

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TABLE 2 Listed scales

Scale	Domain	Referenced Studies in PSP	Validation Outside of PSP
Hasegawa Dementia Rating Scale	Cognitive	Fukui et al.[50]]	AD
Frontal Behavioral Inventory	Cognitive	Borroni et al.,[51] Kertesz and McMonagle[52], Kertesz et al.[53]	Dementing disorders
Wechsler Adult Intelligence Scale	Cognitive	Pillon et al.[54], Milberg and Albert[55]	Dementia, AD
Hospital Anxiety and Depression Scale	Psychiatric	Schrag et al. [56]	PD
Columbia Suicide Severity Rating Scale	Psychiatric	Esmonde et al.[57]	None
SCOPA-AUT	Autonomic	Berganzo et al.[58]	PD
Unified Multiple System Atrophy Scale	Motor	Winter et al.[47], Berganzo et al.[58]	MSA
PDQ-39	QoL	Schrag et al.[31]	PD
Schwab and England Activities of Daily Living	ADLs	Weiner et al.[59]	PD
Hyogo Activities of Daily Living Scale	ADLs	Hirono et al.[60]	None
Yesterday Interview	Quality of Life	Lomax et al.[61]	None
Epworth Sleepiness Scale	Sleep	Gama et al.[62]	PD
Pittsburgh Sleep Quality Index	Sleep	Gama et al.[62]	PD
Restless Legs Scale	Sleep	Gama et al.[62]	RLS
Parkinson Disease Sleep Scale	Sleep	Sixel-Doring et al.[63]	PD
Berlin Questionnaire	Sleep	Gama et al.[62]	

AD, Alzheimer's disease; SCOPA-AUT, Scales for Outcomes in Parkinson's disease–autonomic; PDQ-39, Parkinson Disease Questionnaire-39; QoL, quality of life scale.

severe oculomotor, cognitive, and bulbar function) are not sufficiently addressed by the scale. The UPDRS section III fulfills criteria for Recommended scale for assessment of motor aspects of PSP, excluding ocular function.

FAB

The FAB was designed as a brief tool for assessment of frontal lobe function.[12] It consists of six subtests ranked from 0 (cannot perform) to 3 (no error): conceptualization; mental flexibility; motor programming; sensitivity to interference; inhibitory control; and environmental autonomy. It has been applied in several studies, including PSP clinical trials conducted by multiple researchers (criteria 1 and 2).[21–23] It has been validated and shown to identify frontal lobe dysfunction, including, but not separately, in patients with PSP.[12] Inter-rater reliability of the FAB total score was reported to be "high ($\kappa = 0.87$)"; however, this analysis only included 17 patients, and it is not clear whether a weighted kappa with quadrate weights was used.[12] The FAB discriminates between PSP, MSA and Parkinson's disease (PD), given that mean scores were lower in PSP than in MSA ($P = 0.02$) or PD ($P < 0.001$).[21] The FAB also measures changes during treatment or with disease progression (criterion 3).[21,23] The main strengths of the scale are that it is easy and quick to perform and requires little training. The major weakness is that major motor disability or communication problems can interfere with performance of the test. The FAB fulfils the criteria for Recommended scale to assess the severity of dysexecutive syndromes in PSP, with the limitation that additional validation specifically in PSP and additional inter-rater reliability studies are required.

Suggested Scales

NNIPPS-PPS

The NNIPPS-PPS scale was designed to determine disease progression and severity of patients with PSP and MSA throughout the disease for use in natural history studies and clinical trials.[9] The scale was developed by a consensus of experts who selected items from various scales, including items from the UPDRS,[11] the PSPRS,[8] the International Cooperative Ataxia Rating Scale,[24] the global ataxia score of the Expanded Disability Status Scale,[25] and items from the Autonomic Symptom Profile.[26] Dimensions of the scale include: (1) functional disability (ADLs); (2) mental function (cognition, mood, and behavior); (3) motor disability (rigidity and bradykinesia); (4) tremor; (5) oculomotor function; (6) cerebellar signs; (7) pyramidal signs; (8) dysautonomia; (9) bulbar/pseudobulbar symptoms; (10) myoclonus; and (11) dystonia. The preliminary version was comprised of 109 items and reduced to 85 items because of redundancy or inappropriateness, with a severity ranging from 0 to 6 (normal to very severe), with a majority of items (65) scored on a 5-point scale (0–4). An additional two items were dropped because of missing data and lack of correlation with any of the factors for a total of 83 items. Time to complete the scale is 30 to 45 minutes.

The validity of the 83-item scale was measured in 317 patients with PSP and 358 MSA patients (criterion 1), and inter-rater reliability was measured in 116 patients (PSP = 42; MSA = 74).[9] Principal component analysis extracted 15 factors, which correspond to the predefined expected domains.[9] The internal consistency of the domains were acceptable to high for all the subscores (Cronbach's alpha = 0.68–0.94), except the pyramidal score.[9] Convergent validity was good, as shown by high correlation of the total score with other global severity scales: Clinician Global Impression of disease severity[27] ($\rho = 0.72$); H & Y[28] ($\rho = 0.76$); and Schwab & England Activity Daily Living scale[29] ($\rho = -0.80$).[9] Inter-rater reliability of the total score was almost perfect (ICC = 0.94) and for the subscores were moderate to almost perfect (ICC = 0.73–0.93), except for myoclonus (ICC = 0.54).[9] Responsiveness to change was highly significant, with the exception of the orthostatic, hypotension, myoclonus, and tremor sections. There were no floor or ceiling effects (criterion 3).[9]

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This new scale has the advantage of being able to measure severity and progression of multiple features that characterize not only PSP, but also MSA. The scale has been appropriately validated for its total and subscores in a larger sample, followed prospectively by multiple investigators. The NNIPPS-PPS requires a smaller sample than the PSPRS to detect treatment effects, compared to the PSPRS and the UPDRS, and less training than the PSPRS.[9] Weaknesses of the scale include lack of use by investigators other than those who developed the scale and long administration time. The NNIPPS-PPS is "Suggested" as a rating scale for PSP. Two of the three criteria are met (applied in PSP and successful clinimetric testing). It has not been used by groups other than the development team as yet.

PSP-QoL

The PSP-QoL is a patient-reported outcome measure, specifically designed to assess the quality of life of patients with PSP.[30] There are 45 items and two subscales: physical and mental impact. Items are scored from 0 (no problem) to 4 (extreme problems). The total subscale sum scores are linearly converted into a 0 to 100 scale. The PSP-QoL, with a 4-week time frame, can be completed by the patients or caregiver. The PSP-QoL form includes a visual analog scale (VAS) about the patient's satisfaction with overall life. It takes 30 minutes to complete.

Properties of the PSP-QoL were evaluated in a sample of 225 patients with PSP (criteria 1).[10,30] There are no published studies using the PSP-QoL besides the ones by its developers.[10,30,31] Scaling assumptions have been met,[32] and exploratory factor analysis supported a two-factor structure (physical and mental subscales). Acceptability was within standards,[32] with absence of floor and ceiling effects (0%–3%). Reliability was supported by good internal consistency, with Cronbach's alpha coefficients >0.90 for both subscales[33] and adequate test-retest reliability (ICC >0.90).[32] A panel of experts in movement disorders determined that content validity was good. Internal construct validity was verified through a moderate correlation between the two subscales. Convergent and discriminant validity was supported by correlations between the PSP-QoL subscales and other measures (EuroQol-5D [EQ-5D] and the Hospital Anxiety and Depression Scale), with no differences by gender (criteria 3).[34–36] Sensitivity to change has not been tested.

The PSP-QoL was developed specifically for PSP and displays good psychometric properties. The scale may be rated by the patient or a caregiver, and it covers both physical and mental aspects. Subscale scores are rated in a 0 to 100 metric, which helps interpretation. The scale is available free of charge.[30,31] The major weakness of the PSP-QoL is the low number of studies that have used this scale. In addition, certain mental subscale items assessing nonobservable issues might be difficult to rate by proxy. Further studies are needed to determine responsiveness and inter-rater reliability. The long time it takes to complete the PSP-QoL limits its use in clinical situations, especially for advanced-stage patients, but it is an appropriate scale for research purposes. The PSP-QoL is only available in English.

The PSP-QoL fulfills criteria for Suggested scale to assess the severity of PSP-related problems. To reach the "Recommended" status, it would need to have been used by other researchers besides its developers. The scale shows very good psychometric properties, but replication studies will be useful to confirm the measurement properties of the PSP-QoL and evaluate the scale's responsiveness.

The Neuropsychiatric Inventory

The Neuropsychiatric Inventory (NPI) evaluates the frequency and severity of 10 behaviors observed in individuals with cognitive disorders.[37] For each item, a score of 1 to 4 for frequency and 1 to 3 for severity are assigned. Caregiver distress is also rated (0–5). The scale has been validated in dementing disorders, including mixed populations of caregivers of Alzheimer's disease patients, vascular dementia, and other dementing disorders. It is not clear that PSP was included in these validation studies.[38] There is high content validity for psychopathology and acceptable concurrent validity. Inter-rater reliability ranges from 89% to 100% and test-retest reliability from 0.79 to 0.86. The NPI has been applied in several clinical studies in PSP (criteria 1 and 2).[39,40] Strengths of the scale are quick administration in less than 10 minutes, it explores a wide range of psychopathology, and it utilizes information from the caregiver. The main weaknesses are that some of the behaviors in the scale are uncommon in PSP populations and the diagnostic utility of the scale is not clear. The NPI fulfils criteria for Suggested scale for neuropsychiatric aspects of PSP with the limitation that it still requires specific validation in patients with PSP.

The Mini–Mental State Examination

The Mini–Mental State Examination (MMSE) is a standardized mental status examination testing seven cognitive domains: orientation to time and place; registration and recall of three words; attention and calculation; language; and visual construction.[41] Median administration time is 10 minutes. Although there is widespread use of the MMSE in PSP and other disorders (criteria 1 and 2), clinimetric properties have not been reported. Strengths of the MMSE are its widespread use, quick administration time, and the ability to compare to other cognitive disorders. Weaknesses include proprietary protection, paucity of clinimetrics in PSP, and that it is only a brief screen of cognitive abilities. The MMSE fulfills criteria for Suggested scale to assess cognitive aspects in PSP.

The Dementia Rating Scale

The Dementia Rating Scale (DRS) is a standardized mental status examination with five subsections: attention; initiation/perseveration; construction; conceptualization; and memory.[42] The scale has been used in PSP by groups other than the authors (criteria 1 and 2). Median administration time is

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25 minutes. In studies comparing total DRS scores for PSP and controls, the area under the receiver operating characteristic curve was 0.93,[43] but there are no other clinimetric data in PSP. The DRS subscores can distinguish PSP from other dementias, but not from MSA. In both conditions, initiation and perseveration subscore deficits predominate.[22,44,45] No study has measured item clustering or criterion validity of the DRS in PSP. Strengths of the DRS are its comprehensiveness and its widespread acceptance. Weaknesses include proprietary protection, long administration time, and paucity of clinimetrics in PSP. Motor disability and communication problems may lead to missing or biased data. The DRS fulfills criteria for Suggested scale to assess cognitive aspects in PSP.

H & Y

The H & Y scale is a clinical function scale developed to measure the severity of PD.[28] It is a one-question scale with one to five stages and takes 2 minutes to administer. The strengths of the scale are that it is the most widely and commonly used scale in parkinsonian disorders[46–48] and is weighted heavily toward postural instability assessments, which are impacted early in PSP. However, there are few formal studies of reliability and validity of the scale and no clinimetric studies in PSP. In addition, there is ambiguity in the scale given that it measures both objective signs on examination and impairment of disease on the patient.[46] It only has five options with a large variety of impairment severities collapsed together.[46] This scale fulfills criteria for Suggested scale in PSP for measurement of clinical function.

EQ-5D

The EQ-5D is a generic health-status and quality-of-life measure.[35] It comprises five dimensions (mobility, self-care, usual activities, pain/discomfort, and anxiety/depression), is rated on a 3-point scale (from 1 [no problem] to 3 [severe problem]), that yields a summary index (maximum score: 1, representing better quality of life). There is a VAS, from 0 (worst health status) to 100 (best health status). The strength of the scale is that the EQ-5D is a generic measure and its scores correlate with depression and disease severity and may distinguish between PSP and MSA at presentation (PSP patients present with significantly higher scores; criteria 1 and 2),[10,47] but there is only limited information about the other psychometric properties in PSP. The main disadvantage is that, as a generic instrument, it underestimates health problems in PSP owing to the fact that it does not incorporate many PSP aspects, such as balance, falls, or social impairments.[31,49] This scale fulfills criteria for Suggested scale in PSP because it has adequate psychometric properties for a generic health-related quality-of-life instrument in PSP, but requires further psychometric testing in PSP.

Conclusions and Recommendations

Three scales are Recommended for use in PSP; however, only one covers the global features of disease. The other two scales focus on motor and cognitive deficits. Only one of the three PSP- specific scales (PSPRS) can currently be recommended for use given that two recently developed scales, the NNIPPS-PPS and PSP-QoL, have not been used by investigators outside of the development team. A benefit of these PSP-specific scales is that they are more comprehensive in measuring the PSP phenotype. Despite our recommendation, the PSPRS has several clinimetric measures that need to be studied further, including content validity and scaling acceptability.

For more in-depth investigation into particular or associated symptoms in PSP, only the UPDRS section III and FAB are recommended for use, but both need further validation in PSP. In general, a rating scale that encompasses the major nonmotor features in PSP is needed, similar to part 1 of the MDS-UPDRS. Assessment of the nonmotor features is recognized to be of growing importance and further scales to assess these in depth should be validated.

A major weakness of the scales reviewed is that many are available in English only. This project was also limited by a lack of information regarding the clinimetric testing of several of the scales. Methodological problems, including missing values, were not available for many of the clinimetric studies. Some of the scales, where noted, were tested in heterogenous populations, which included patients with PSP or other diseases. These issues could have resulted in a higher rating from the task force than warranted.

In addition, further studies should explore the responsiveness of the scales and the minimally clinically significant change given that this was infrequently addressed. At the present time, we do not recommend the development of new scales for clinical features in PSP until the NNIPPS-PPS and PSP-QoL are evaluated more widely.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

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References

1. Steele JC, Richardson JC, Olszewski J. Progressive supranuclear palsy. A heterogeneous degeneration involving the brain stem, Basal Ganglia and cerebellum with vertical gaze and pseudobulbar palsy, nuchal dystonia and dementia. *Arch Neurol* 1964;10:333–359.
2. Litvan I, Agid Y, Calne D, et al. Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report of the NINDS-SPSP international workshop. *Neurology* 1996;47:1–9.
3. Schrag A, Barone P, Brown RG, et al. Depression rating scales in Parkinson's disease: critique and recommendations. *Mov Disord* 2007;22:1077–1092.
4. Fernandez HH, Aarsland D, Fenelon G, et al. Scales to assess psychosis in Parkinson's disease: critique and recommendations. *Mov Disord* 2008;23:484–500.
5. Colosimo C, Martinez-Martin P, Fabbrini G, et al. Task force report on scales to assess dyskinesia in Parkinson's disease: critique and recommendations. *Mov Disord* 2010;25:1131–1142.
6. Martinez-Martin P, Forjaz MJ. *How to Evaluate Validation Data*. Oxford: Oxford University Press; 2012.
7. Terwee CB, Bot SD, de Boer MR, et al. Quality criteria were proposed for measurement properties of health status questionnaires. *J Clin Epidemiol* 2007;60:34–42.
8. Golbe LI, Ohman-Strickland PA. A clinical rating scale for progressive supranuclear palsy. *Brain* 2007;130:1552–1565.
9. Payan CA, Viallet F, Landwehrmeyer BG, et al. Disease severity and progression in progressive supranuclear palsy and multiple system atrophy: validation of the NNIPPS–Parkinson Plus Scale. *PLoS One* 2011;6:e22293.
10. Schrag A, Selai C, Quinn N, Hobart J, Lees A, Litvan I. Measuring quality of life in progressive supranuclear palsy. In: Jenkinson C, Peters M, Bromberg M, eds. *Quality of Life Measurement in Neurodegenerative and Related Conditions*. Cambridge: Cambridge University Press; 2011:52–59.

doi:10.1002/mdc3.12130

11. Fahn S, Elton R; Committee MotUD. Unified Parkinson's Disease Rating Scale. In: Fahn S, Marsden C, Calne D, Goldstein M, eds. *Recent Developments in Parkinson's Disease*. Florham Park: MacMillan Healthcare Information; 1987:153–163.

12. Dubois B, Slachevsky A, Litvan I, Pillon B. The FAB: a frontal assessment battery at bedside. *Neurology* 2000;55:1621–1626.

13. Stamelou M, Reuss A, Pilatus U, et al. Short-term effects of coenzyme Q10 in progressive supranuclear palsy: a randomized, placebo-controlled trial. *Mov Disord* 2008;23:942–949.

14. Lindemann U, Nicolai S, Beische D, et al. Clinical and dual-tasking aspects in frequent and infrequent fallers with progressive supranuclear palsy. *Mov Disord* 2010;25:1040–1046.

15. Chiu WZ, Kaat LD, Seelaar H, et al. Survival in progressive supranuclear palsy and frontotemporal dementia. *J Neurol Neurosurg Psychiatry* 2010;81:441–445.

16. Golbe LI, Davis PH, Schoenberg BS, Duvoisin RC. Prevalence and natural history of progressive supranuclear palsy. *Neurology* 1988;38:1031–1034.

17. Stebbins GT, Goetz CG. Factor structure of the Unified Parkinson's Disease Rating Scale: motor examination section. *Mov Disord* 1998;13:633–636.

18. Stebbins GT, Goetz CG, Lang AE, Cubo E. Factor analysis of the motor section of the unified Parkinson's disease rating scale during the off-state. *Mov Disord* 1999;14:585–589.

19. Kroonenberg PM, Oort FJ, Stebbins GT, Leurgans SE, Cubo E, Goetz CG. Motor function in Parkinson's disease and supranuclear palsy: simultaneous factor analysis of a clinical scale in several populations. *BMC Med Res Methodol* 2006;6:26.

20. Cubo E, Stebbins GT, Golbe LI, et al. Application of the Unified Parkinson's Disease Rating Scale in progressive supranuclear palsy: factor analysis of the motor scale. *Mov Disord* 2000;15:276–279.

21. Paviour DC, Price SL, Lees AJ, Fox NC. MRI derived brain atrophy in PSP and MSA-P. Determining sample size to detect treatment effects. *J Neurol* 2007;254:478–481.

22. Brown RG, Lacomblez L, Landwehrmeyer BG, et al. Cognitive impairment in patients with multiple system atrophy and progressive supranuclear palsy. *Brain* 2010;133:2382–2393.

23. Whitwell JL, Xu J, Mandrekar JN, Gunter JL, Jack CR, Jr., Josephs KA. Rates of brain atrophy and clinical decline over 6 and 12-month intervals in PSP: determining sample size for treatment trials. *Parkinsonism Relat Disord* 2011;18(3):252–256.

24. Trouillas P, Takayanagi T, Hallett M, et al. International Cooperative Ataxia Rating Scale for pharmacological assessment of the cerebellar syndrome. The Ataxia Neuropharmacology Committee of the World Federation of Neurology. *J Neurol Sci* 1997;145:205–211.

25. Kurtzke JF. Rating neurologic impairment in multiple sclerosis: an expanded disability status scale (EDSS). *Neurology* 1983;33:1444–1452.

26. Suarez GA, Opfer-Gehrking TL, Offord KP, Atkinson EJ, O'Brien PC, Low PA. The autonomic symptom profile: a new instrument to assess autonomic symptoms. *Neurology* 1999;52:523–528.

27. Nunnally J, Bernstein I. *Psychometric Theory*. New York: McGraw-Hill press; 1994.

28. Hoehn MM, Yahr MD. Parkinsonism: onset, progression and mortality. *Neurology* 1967;17:427–442.

29. Schwab R, England A. Projection techniques for evaluating surgery in Parkinson's disease. In: Gillingham F, Donaldson I, eds. *Third Symposium on Parkinson's Disease, Royal College of Surgeons*. Edinburgh: Livingstone; 1969:152–158.

30. Schrag A, Selai C, Quinn N, et al. Measuring quality of life in PSP: the PSP-QoL. *Neurology* 2006;67:39–44.

31. Schrag A, Selai C, Davis J, Lees AJ, Jahanshahi M, Quinn N. Health-related quality of life in patients with progressive supranuclear palsy. *Mov Disord* 2003;18:1464–1469.

32. Ware JE, Jr., Gandek B. Methods for testing data quality, scaling assumptions, and reliability: the IQOLA Project approach. International Quality of Life Assessment. *J Clin Epidemiol* 1998;51:945–952.

33. Trust SACotMO. Assessing health status and quality-of-life instruments: attributes and review criteria. *Qual Life Res* 2002;11:193–205.

34. Peto V, Jenkinson C, Fitzpatrick R. PDQ-39: a review of the development, validation and application of a Parkinson's disease quality of life questionnaire and its associated measures. *J Neurol* 1998;245(Suppl 1): S10–S14.

35. Group E. EuroQol–a new facility for the measurement of health-related quality of life. The EuroQol Group. *Health Policy* 1990;16:199–208.

36. Zigmond AS, Snaith RP. The hospital anxiety and depression scale. *Acta Psychiatr Scand* 1983;67:361–370.

37. Cummings JL, Mega M, Gray K, Rosenberg-Thompson S, Carusi DA, Gornbein J. The Neuropsychiatric Inventory: comprehensive assessment of psychopathology in dementia. *Neurology* 1994;44:2308–2314.

38. Cummings JL. The Neuropsychiatric Inventory: assessing psychopathology in dementia patients. *Neurology* 1997;48:S10–S16.

39. Aarsland D, Litvan I, Larsen JP. Neuropsychiatric symptoms of patients with progressive supranuclear palsy and Parkinson's disease. *J Neuropsychiatry Clin Neurosci* 2001;13:42–49.

40. Kulisevsky J, Litvan I, Berthier ML, Pascual-Sedano B, Paulsen JS, Cummings JL. Neuropsychiatric assessment of Gilles de la Tourette patients: comparative study with other hyperkinetic and hypokinetic movement disorders. *Mov Disord* 2001;16:1098–1104.

41. Folstein MF, Folstein SE, McHugh PR. "Mini-mental state". A practical method for grading the cognitive state of patients for the clinician. *J Psychiatr Res* 1975;12:189–198.

42. Mattis S. Dementia rating scale: professional manual. Odessa 1998.

43. Bak TH, Crawford LM, Hearn VC, Mathuranath PS, Hodges JR. Subcortical dementia revisited: similarities and differences in cognitive function between progressive supranuclear palsy (PSP), corticobasal degeneration (CBD) and multiple system atrophy (MSA). *Neurocase* 2005;11:268–273.

44. Aarsland D, Litvan I, Salmon D, Galasko D, Wentzel-Larsen T, Larsen JP. Performance on the dementia rating scale in Parkinson's disease with dementia and dementia with Lewy bodies: comparison with progressive supranuclear palsy and Alzheimer's disease. *J Neurol Neurosurg Psychiatry* 2003;74:1215–1220.

45. Rosser AE, Hodges JR. The Dementia Rating Scale in Alzheimer's disease, Huntington's disease and progressive supranuclear palsy. *J Neurol* 1994;241:531–536.

46. Goetz CG, Poewe W, Rascol O, et al. Movement Disorder Society Task Force report on the Hoehn and Yahr staging scale: status and recommendations. *Mov Disord* 2004;19:1020–1028.

47. Winter Y, Spottke AE, Stamelou M, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegener Dis* 2011;8:438–446.

48. Goetz CG, Leurgans S, Lang A, Litvan I. Progression of gait, speech and swallowing deficits in progressive supranuclear palsy. *Neurology* 2003;60:917–922.

49. Schrag A, Selai C, Jahanshahi M, Quinn NP. The EQ-5D–a generic quality of life measure-is a useful instrument to measure quality of life in patients with Parkinson's disease. *J Neurol Neurosurg Psychiatry* 2000;69:67–73.

50. Fukui T, Yamazaki T, Kinno R. Can the 'head-turning sign' be a clinical marker of Alzheimer's disease? *Dement Geriatr Cogn Dis Extra* 2011;1:310–317.

51. Borroni B, Alberici A, Agosti C, Cosseddu M, Padovani A. Pattern of behavioral disturbances in corticobasal degeneration syndrome and progressive supranuclear palsy. *Int Psychogeriatr* 2009;21:463–468.

52. Kertesz A, McMonagle P. Behavior and cognition in corticobasal degeneration and progressive supranuclear palsy. *J Neurol Sci* 2010;289:138–143.

53. Kertesz A, McMonagle P, Jesso S. Extrapyramidal syndromes in frontotemporal degeneration. *J Mol Neurosci* 2011;45:336–342.

54. Pillon B, Gouider-Khouja N, Deweer B, et al. Neuropsychological pattern of striatonigral degeneration: comparison with Parkinson's disease and progressive supranuclear palsy. *J Neurol Neurosurg Psychiatry* 1995;58:174–179.

55. Milberg W, Albert M. Cognitive differences between patients with progressive supranuclear palsy and Alzheimer's disease. *J Clin Exp Neuropsychol* 1989;11:605–614.

56. Schrag A, Sheikh S, Quinn NP, et al. A comparison of depression, anxiety, and health status in patients with progressive supranuclear palsy and multiple system atrophy. *Mov Disord* 2010;25:1077–1081.

57. Esmonde T, Giles E, Gibson M, Hodges JR. Neuropsychological performance, disease severity, and depression in progressive supranuclear palsy. *J Neurol* 1996;243:638–643.

doi:10.1002/mdc3.12130

58. Berganzo K, Tijero B, Somme JH, et al. SCOPA-AUT scale in different parkinsonisms and its correlation with (123) I-MIBG cardiac scintigraphy. *Parkinsonism Relat Disord* 2012;18:45–48.

59. Weiner WJ, Minagar A, Shulman LM. Pramipexole in progressive supranuclear palsy. *Neurology* 1999;52:873–874.

60. Hirono N, Kobayashi H, Mori E. [Caregiver burden in dementia: evaluation with a Japanese version of the Zarit caregiver burden interview]. *No To Shinkei* 1998;50:561–567.

61. Lomax CL, Brown RG, Howard RJ. Measuring disability in patients with neurodegenerative disease using the 'Yesterday Interview'. *Int J Geriatr Psychiatry* 2004;19:1058–1064.

62. Gama RL, Tavora DG, Bomfim RC, Silva CE, de Bruin VM, de Bruin PF. Sleep disturbances and brain MRI morphometry in Parkinson's disease, multiple system atrophy and progressive supranuclear palsy – a comparative study. *Parkinsonism Relat Disord* 2010;16:275–279.

63. Sixel-Doring F, Schweitzer M, Mollenhauer B, Trenkwalder C. Polysomnographic findings, video-based sleep analysis and sleep perception in progressive supranuclear palsy. *Sleep Med* 2009;10:407–415.

Supporting Information

Additional Supporting Information may be found in the online version of this article:

Data S1. The supplementary material contains additional information regarding the scales in this report. Included is a description of each scale, scale properties, use, clinimetrics, and overall impression for both suggested and recommended scales.

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