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Medical decision-making in progressive supranuclear palsy: A comparison to other neurodegenerative disorders

Adam Gerstenecker[a,b,c,*], Lacey Grimsley[a], Brittney Otruba[e], Linda Cowden[a], Daniel C. Marson[a,b], Kristen Triebel Gerstenecker[a,b,c], Roy C. Martin[a,b,c], Erik D. Roberson[a,b,c,d]

[a] Department of Neurology, University of Alabama at Birmingham, Birmingham, AL, USA
[b] Alzheimer's Disease Center, University of Alabama at Birmingham, Birmingham, AL, USA
[c] Evelyn F. McKnight Brain Institute, University of Alabama at Birmingham, Birmingham, AL, USA
[d] Center for Neurodegeneration and Experimental Therapeutics, University of Alabama at Birmingham, Birmingham, AL, USA
[e] Department of Psychiatry, University of Alabama at Birmingham, Birmingham, AL, USA

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ABSTRACT

Introduction: Cognitive dysfunction is common in progressive supranuclear palsy (PSP) but its effect on medical decision-making has not been well studied. To address this gap in the research literature, we compared the medical decision-making capacity of patients with PSP to groups of patients with other neurodegenerative disorders. We also investigated the cognitive correlates of medical decision-making in our PSP sample.
Methods: The sample was composed of 65 participants that belonged to five study groups: 13 with PSP, 13 with PD-MCI, 13 with PDD, 13 with AD, and 13 cognitively healthy controls. A neuropsychological battery, including a performance-based measure of medical decision-making capacity, was administered to all study participants.
Results: Over 80% of the PSP group exhibited impairment in some aspect of medical decision-making and rates of impairment in PSP were similar to PDD. A number of cognitive abilities contributed to medical decision-making impairment in the PSP group, with executive function and attention being primary contributors. Medical decision-making was not associated with either disease duration or severity.
Conclusions: Impaired medical decision-making appears to be a prevalent feature of PSP and impairments in specific cognitive domains may negatively affect the ability of this cohort to make sound medical decisions. The cognition of patients with PSP has likely declined to the point of affecting decision-making ability at the time of diagnosis and recruitment to research studies. This finding has important implications about the ability of clinicians and researchers to consent patients with PSP for research or treatment.

1. Introduction

Medical decision-making is a higher-order functional skill that refers to the ability to make sound decisions about medical care and treatment. Medical decision-making is often analyzed using four consent standards derived from the medical and legal literature: *Expressing* a treatment choice, *appreciating* the risks/benefits of a choice, logical *reasoning* about choices, and *understanding* the disease and treatment options [1]. Of these standards, *reasoning* and *understanding* appear to be the most cognitively demanding [2]. In a medical setting, *reasoning* refers to the ability of a patient to consider information in a logical manner, allowing him/her to form a valid judgment or conclusion about diagnosis, prognosis, and treatment options. *Understanding* allows the patient to comprehend and recall factual information about treatment options and make informed medical decisions.

Our group has examined medical decision-making in multiple samples of patients with a neurocognitive disorder and found that level of cognitive decline correlates with impairment in medical decision-making [2]. Deficits in medical decision-making have been noted in patients with mild cognitive impairment (MCI) due to Parkinson's disease (PD) and dementia due to PD [3]. Deficits in medical decision-making have also been noted in Alzheimer's disease (AD), with deficits in verbal memory, executive function, and processing speed being predictive of level of medical decision-making impairment [4].

Progressive supranuclear palsy (PSP) is a rare neurodegenerative, parkinsonian disorder with a median age of onset of 63 years and

* Corresponding author. University of Alabama at Birmingham (UAB) Department of Neurology, 650 Sparks Center, 1720 7th Avenue South, Birmingham, AL, 35294, USA.
E-mail address: atgers@uabmc.edu (A. Gerstenecker).

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disease duration of 7.4 years. Individuals with the PSP-Richardson syndrome (PSP-RS) phenotype present with postural instability and falls, parkinsonism not responding to dopaminergic therapy, and slowing of vertical saccades [5]. Although PSP is most often characterized as a movement disorder, cognitive dysfunction, behavioral disturbances, and functional deficits are common [6–9]. To date, however, no studies have examined the ability of patients with PSP to make medical decisions.

Medical decision-making appears to be an under-investigated topic in PSP. We address this gap in the research literature by administering a vignette-based measure of medical decision-making and a number of neuropsychological measures to a sample of PSP patients and comparing their performance to that of a group of healthy older adults and groups of patients with mild cognitive impairment due to PD (PD-MCI), Parkinson's disease dementia (PDD), and mild AD. Cognitive correlates of medical decision-making were investigated in the PSP sample. Given the level of cognitive impairment in PSP [6,10,11], we hypothesized that the PSP sample would demonstrate decline in *reasoning* and *understanding* and exhibit overall performance similar to that of patients with PDD. Since deficits in executive function are the most pervasive cognitive impairment in PSP [10], we hypothesized that executive dysfunction would be a key neurocognitive predictor of medical decision-making.

2. Methods

2.1. Participants

In total, 65 participants were recruited for studies taking place at the University of Alabama at Birmingham (UAB). All study participants composed one of five groups: 13 with PSP, 13 with PD-MCI, 13 with PDD, 13 with mild AD, and 13 cognitively healthy controls.

All patients with PSP met the NINDS-SPSP criteria [12] for clinically possible or probable PSP, which correspond to the recently published probable MDS-PSP criteria [13]. All patients in the current sample best corresponded to the PSP Richardson's syndrome subtype.

All patients with PD were clinically characterized as having idiopathic PD and met 2015 MDS-PD criteria [14]. Diagnoses of PDD were based upon a clinical history of (1) movement disorder for at least one year prior to onset of cognitive impairment, (2) neurological findings supporting idiopathic PD, (3) neuropsychological test results indicating cognitive impairment in at least two cognitive domains ($\geq$1.5 SD below normal age-matched means), and (4) collateral report of a family member or caregiver for evidence of significant decline in everyday function from premorbid levels due to cognitive decline.

The PD-MCI group had (1) a clinical history of movement disorder for at least one year prior to onset of cognitive impairment, (2) neurological findings supporting idiopathic PD, (3) neuropsychological test results indicating impairment ($\geq$1.5 SD below normal age-matched means) in at least one cognitive domain, and (4) collateral report of a family member or caregiver that the patient had showed cognitive decline but no more than minimal change in higher order activities of daily living from premorbid baseline. These criteria are consistent with prior published criteria [15].

All participants in the AD group were considered to have mild AD dementia. Diagnoses of mild AD were based on published criteria at the time of evaluation, namely the National Institute of Neurological and Communicative Diseases and Stroke/Alzheimer's Disease and Related Disorders Association (NINCDS/ADRDA) criteria for probable AD [16]. These criteria are as follows: memory impairment established by neuropsychological testing, deficits ($\leq$5th percentile using age- and education-corrected scores) in two or more areas of cognition, evidence of continued decline from a previous level of functioning through a collateral source and structured clinical examination or assessment of activities of daily living, no disturbance of consciousness, onset between ages 40 and 90, and the absence of systemic disorders or other brain diseases that could account for the progressive deficits in memory and cognition.

General exclusion criteria for the patient groups included history of major psychiatric disorder (including major depression, but excluding minor depression and dysthymia), history of substance abuse, prior neurosurgical intervention, concomitant medical illness adversely affecting cognition (e.g., obstructive pulmonary disease), metabolic disease, head injury, stroke, or neuro-developmental condition. Patients with clinical features consistent with Dementia with Lewy Bodies (using the one-year rule) or Alzheimer's disease and secondary Parkinsonism were also excluded.

Cognitively normal older adult participants recruited for the current study were required to meet the following criteria: 1) absence of impairment on measures of neurocognitive function, 2) absence of diseases or conditions that could potentially affect cognition, including psychiatric disorder (except mild depression), substance abuse, cerebrovascular disease, or other neurologic diseases; 3) absence of findings on physical examination suggestive of problems with cognition; and 4) absence of the use of medications known to affect cognition. Thus, normal controls were believed to represent a sample comparable to adults living independently in the community.

Diagnoses of PSP, PD-MCI, PDD, AD, and normal control were made by the study's diagnostic consensus conference team, which consisted of neurologists, neuropsychologists, and nursing staff.

All study procedures were approved by UAB's institutional review board and informed consent was obtained for all study participants.

2.2. Procedures

After informed consent, a detailed history was obtained for each participant. For the PSP and PD groups, a neurological examination including the PSP Rating Scale (PSPRS) [17] and Unified Parkinson's Disease Rating Scale (UPDRS) [18], respectively, was also conducted. A baseline neuropsychological evaluation—including a measure of medical decision-making—was administered by a psychological technician who had received training on the battery from a neuropsychologist. Accuracy of the evaluations was checked by a neuropsychologist.

2.3. Measures

Medical Decision-Making Capacity: *understanding*, *reasoning*, and Total score from vignette B of the Capacity to Consent to Treatment Instrument (CCTI) [19].

Cognitive Functioning: Dementia Rating Scale – Second Edition (DRS-2) Attention [20] for attention; Hopkins Verbal Learning Test – Revised [21] (HVLT-R) for verbal learning and memory (Learning Trials Total and Delayed Recall); 10/36 Spatial Recall Test [22,23] for visuospatial learning and memory (Immediate and Delayed Recall); semantic fluency [24] for verbal fluency (Animals, Fruits and Vegetables, Clothing); DRS-2 Construction for visuospatial ability; phonemic/letter fluency [24] (“F” “A” “S”), Oral Trail Making Test Part B [25] (OTMT-B), and Frontal Assessment Battery [26] (FAB) for executive function; and OTMT-A and Oral Symbol Digit Modalities Test [27] for processing speed.

Disease Severity: PSPRS [17] and UPDRS [18].

2.4. Data analyses

First, means and standard deviations or frequency counts were calculated for demographic, medical decision-making, and cognitive data. Second, one-way ANOVAs or Pearson's chi-square tests were conducted to determine if the study groups differed on demographic, medical decision-making, or cognitive data. Tukey's honest significance post hoc tests were utilized to evaluate for group differences if main effects were found for the ANOVAs. Due to CCTI *reasoning* scores being relatively truncated, non-parametric alternatives were used to

Table 1
Group demographics, *Reasoning*, *Understanding*, and general cognition.

	PSP	PD-MCI	PDD	AD	NC	$F/\chi^2/H$	*p*
Demographic							
Age	67 (5.0)	67 (8.5)	68 (4.7)	74 (5.8)	66 (5.0)	3.9	0.007*
Education	15 (2.6)	15 (2.0)	16 (3.0)	13 (2.4)	16 (3.0)	2.0	0.100
Sex	11, 2	10, 3	12, 1	8, 5	11, 2	4.4	0.352
Race	13, 0	13, 0	13, 0	12, 1	11, 2	5.6	0.232
Medical Decision-Making							
Reasoning	3.3 (2.2)	5.0 (3.2)	2.4 (2.1)	2.7 (2.2)	4.9 (2.9)	10.6	0.032*
Understanding	18.9 (11.1)	29.7 (5.8)	19.5 (8.5)	16.4 (5.6)	33.4 (4.7)	13.0	< 0.001*
General Cognition							
DRS-2 Total	117.7 (14.8)	135.3 (4.6)	117.2 (13.9)	120.0 (8.8)	138.9 (3.1)	13.6	< 0.001*

Note. For age, education, Medical Decision-Making, and DRS-2 Total, cells include mean (standard deviation). For sex and race, cells include male, female and Caucasian, African-American, respectively. $F/\chi^2/H$ = analysis of variance, chi-square, or Kruskal-Wallis test statistic, *p* = p value. * indicates significant result. PSP = progressive supranuclear palsy, PD-MCI = Parkinson's disease mild cognitive impairment, PDD = Parkinson's disease dementia, AD = Alzheimer's disease, NC = normal control, DRS-2 = Dementia Rating Scale – Second Edition.

investigate for possible differences on this consent standard. Third, z-scores for CCTI and neuropsychological variables were calculated based on the available normative means and standard deviations [28]. When there was more than one cognitive measure administered for a domain, an average z-score for the combined measures was used to represent an aggregated domain score. Fourth, for the PSP group, Pearson product moment correlations were calculated to examine the relationship among CCTI variables and neurocognitive performances, age, education, disease duration, and PSPRS scores. Finally, variables that were found to be significantly associated with a particular CCTI variable (i.e., CCTI Total, *understanding*, or *reasoning*) were then used to construct three forced-entry, linear regression models.

Due to the limited sample size of the PSP group and associated limited statistical power, a two-tailed .05 level of significance was used for all analyses to balance the probability of type I and type II errors.

3. Results

3.1. Sample characteristics

Table 1 lists demographics and DRS-2 Total score for the study groups.

The study groups were predominantly Caucasian, male, and well educated. Age was the only demographic variable that differed between the study groups. This difference was due to the AD group being older than the PSP ($p = 0.021$), PD-MCI ($p = 0.030$), and normal control ($p = 0.009$) groups.

For the PSP group, average disease duration was 3.3 (standard deviation 2.0) years and average PSPRS Total score was 37.5 (8.7). The UPDRS Motor subsection was administered to the PD groups. UPDRS Motor score was 9.9 (4.9) and 13.2 (4.0) for the PD-MCI and PDD groups, respectively.

A main effect for general cognition (DRS-2 Total score) was observed. Both the normal control and PD-MCI groups obtained higher DRS-2 Total scores than the PSP ($p < 0.001$), PDD ($p < 0.001$), and AD ($p < 0.001$ and $p = 0.003$, respectively) groups.

3.2. medical decision-making

Table 1 also lists group performance on the CCTI consent standards of *reasoning* and *understanding*.

Despite a main effect for *reasoning* being found, differences were not noted at the group level. However, when considering effect size (Cohen's *d*), medium or large effects were noted for several comparisons: normal control versus PSP ($d = 0.62$), PDD ($d = 0.99$), and AD ($d = 0.85$); PSP versus PD-MCI ($d = 0.62$); and PD-MCI versus PDD ($d = 0.96$), and AD ($d = 0.84$).

A main effect for CCTI *understanding* was noted. Both the normal control and PD-MCI groups obtained higher scores on the CCTI *understanding* consent standard than the PSP ($p < 0.001$ and $p = 0.004$, respectively), PDD ($p < 0.001$ and $p = 0.008$, respectively), and AD ($p < 0.001$) groups.

3.3. Medical decision-making impairment for the PSP group

The number (percent) impaired of patients in the PSP group that showed impairment on CCTI variables were as follows: 11 (84.6%) for CCTI Total, 4 (30.8%) for *reasoning*, and 10 (76.9%) for *understanding* (Table 2).

3.4. Correlates of medical decision-making for the PSP group

None of age, education, disease duration, or PSPRS Total score were significantly associated with any CCTI variable in the PSP group (Table 3). Although consistent with our previous investigations of associations between cognition and clinical factors in PSP [10], the lack of significant correlations between disease duration,PSPRS Total score and medical decision-making remains interesting. Given that PSP is rarely diagnosed early in its course, these findings may indicate that most PSP patients recruited to clinical research studies already experience significant motor and cognitive impairment. In addition, as most measures of disease severity in PSP are primarily motor-based, it could be that neural systems involved in motor and cognitive function are differentially affected in PSP.

A number of cognitive variables were associated with medical decision-making in the PSP group. CCTI Total was significantly associated with Executive Function ($r = 0.70$, $p = 0.008$) and Attention ($r = 0.61$, $p = 0.027$). The association between *reasoning* and Attention was also significant ($r = 0.65$, $p = 0.016$). *Understanding* was significantly correlated with Executive Function ($r = 0.78$, $p = 0.002$), Attention ($r = 0.69$, $p = 0.009$), Processing Speed ($r = 0.62$, $p = 0.024$), and Verbal Fluency ($r = 0.57$, $p = 0.043$).

We next used cognitive variables that were significantly associated with CCTI Total, *reasoning*, and *understanding* in the PSP group to construct three regression models (Table 4). For CCTI Total, *reasoning*,

Table 2
CCTI impairment ratings for the PSP group.

CCTI Variable	No Impairment	Mild/Moderate Impairment	Severe Impairment
CCTI Total	2 (15.4)	2 (15.4)	9 (69.2)
Reasoning	9 (69.2)	3 (23.1)	1 (7.7)
Understanding	3 (23.1)	2 (15.4)	8 (61.5)

Note. Values are *n* (%). PSP = progressive supranuclear palsy, CCTI = Capacity to Consent to Treatment Instrument.

Table 3
CCTI clinical correlates for the PSP group.

Clinical Variable	CCTI Total	*Reasoning*	*Understanding*
Disease Duration	0.046	0.327	0.236
PSPRS Total	−0.091	0.290	−0.104
Age	−0.153	−0.058	−0.159
Education	0.193	0.286	0.335

Note. Values are Pearson product moment correlations. PSP = progressive supranuclear palsy, CCTI = Capacity to Consent to Treatment Instrument.

Table 4
Results of the linear regression models for the PSP group.

CCTI Variable	F; *p*	R^2	SEE	β, SE
CCTI Total	5.5; 0.025	0.52	10.9	
Executive Function				1.4, 1.4
Attention				6.5, 3.3
Constant				37.4, 8.1
Reasoning	3.1; 0.104	0.47	3.3	
Attention				1.4, 0.8
Constant				3.2, 1.0
Understanding	2.3; 0.146	0.54	9.2	
Attention				5.5, 3.2
Verbal Fluency				1.6, 3.3
Executive Function				1.2, 2.0
Processing Speed				−2.7, 4.8
Constant				28.5, 7.9

Note. PSP = progressive supranuclear palsy; SEE = standard error of the estimate of the regression model; β = unstandardized beta weights; SE = standard error of coefficient; CCTI = Capacity to Consent to Treatment Instrument.

and *understanding*, individual models accounted for 52%, 47%, and 54% of shared variance, respectively.

4. Discussion

In this paper, we investigated aspects of medical decision-making in PSP. Ensuring that a patient adequately comprehends medical and treatment information is essential from an ethical and legal standpoint. However, medical decision-making has been shown to be impaired in a number of samples of patients with a neurologic condition [2]. To our knowledge, the current study is the first to examine medical decision-making in a PSP sample using a performance-based capacity measure. Our results indicate that well over half of PSP patients may be impaired in some aspect of medical decision-making and that rates of medical decision-making impairment in PSP are similar to PDD. A number of cognitive variables were associated with medical decision-making in our PSP sample, with executive function and attention being identified as primary predictors. However, medical decision-making was not significantly associated with either disease duration or severity, which likely indicates that cognitive decline has progressed to the point of affecting decision-making at the time of diagnosis of PSP and recruitment into research studies. This finding raises questions about the ability of patients with PSP to adequately consent to research and treatment at the time of initial diagnosis.

As expected, the PSP group performed poorly on all CCTI variables examined in the present study. The ability of the PSP group to comprehend presented medical information (i.e., CCTI *understanding*) was significantly poorer than that of the normal control and PD-MCI groups but statistically similar to that of the PDD and AD groups. When considering effect size, the same pattern was observed for the ability of the PSP group to logically consider presented medical information (i.e., CCTI *reasoning)*.

Rates of medical decision-making impairment were high for the PSP group and similar to that of PDD. For CCTI Total, 85% of the sample exhibited impaired performance. For the consent standards of *reasoning*, and *understanding*, impairment was present in 31% and 77%, respectively (Table 2). When medical decision-making was compared in the PSP group to the other patient groups, it was striking how closely rates of impairment matched that of PDD (i.e., 31% and 81% in PDD compared to 31% and 77% in PSP for *reasoning* and *understanding*, respectively) [3]. However, the concordance between rates of medical decision-making impairment between the groups was not unexpected given that DRS-2 scores were virtually identical between the PSP and PDD groups. In relation to AD, the PSP group demonstrated less medical decision-making impairment than the mild AD group (i.e., 52% and 100% in mild AD for *reasoning* and *understanding*, respectively) [29]. Given that scores on a measure of general cognition (i.e., DRS-2) were similar between the PSP and AD samples, the finding that medical decision-making is more impaired in AD likely highlights the importance of semantic ability in making treatment decisions, as loss of semantic memory and ability is pervasive in AD.

Identifying the specific neuropsychological abilities related to medical decision-making could lead to a better ability of clinicians to screen patients for decision-making impairment, as such findings could be used as “red flags” to indicate that diminished decision-making capacity may be present. In our PSP sample, CCTI Total, *reasoning*, and *understanding* were significantly associated with a number of cognitive variables. For CCTI Total, z-scores for Executive Function and Attention accounted for 52% of shared variance. For *reasoning*, the z-score for Attention accounted for 47% of shared variance. For *understanding*, z-scores for Attention, Verbal Fluency, Executive Function, and Processing Speed accounted for 54% of shared variance (Table 4). Thus, measures tapping the aforementioned cognitive domains may represent an opportunity for clinicians to quickly screen for medical decision-making impairment in PSP. For example, the measures included in the model for CCTI Total took ~17 min to administer. For *reasoning* and *understanding*, the included measures took ~7 min and ~23 min to administer, respectively.

Although the above administration times may be impractical for standard clinical visits, the findings of the current study have clinical and research implications. Our finding that over 80% of the PSP group had difficulty making sound treatment decisions suggests that medical decision-making should be considered by clinicians when working with these patients. Although effective pharmacological therapies for PSP do not currently exist, opportunities for participation in clinical trials are available. However, our findings suggest that most patients with PSP likely lack the comprehension and reasoning skills necessary to make informed decisions about the potential costs versus benefits of the experimental treatments used in clinical trials. Accordingly, clinicians enrolling PSP patients in clinical trials are encouraged to give careful attention to the informed consent process because 2004 Alzheimer's Association guidelines stipulate that a formal health care agent (HCA) must be appointed and present during the consent process for patients with significant medical decision-making impairments [30]. Clinical recommendations include simplifying language used in verbal dialogue and written consent forms, presenting information in manageable segments, asking patients to explain the information presented to them, querying patients to assess their understanding of the treatment information presented, and/or using multiple modalities (e.g., auditory and visual) to present treatment information.

There are some limitations and future directions to be considered. First, all patients composing the current sample met NINDS-SPSP criteria for probable or possible PSP. These criteria identify PSP-RS well but are limited in the identification of other PSP phenotypes. Future studies should classify patients according to their subtype as the methodology for distinguishing between subtypes improve. Second, the small sample size in combination with the number of potential independent variables increases the risk of the results not being replicated in future samples. However, the potential for false negatives may represent a larger risk in the current small sample. Third, the current sample was majority male and Caucasian. More diverse samples should

be examined in future studies. Finally, multiple measures were used to form domain z-scores. It is possible that the number of measures could be shortened to develop a brief screen for medical decision-making impairment in PSP. Despite these limitations, these preliminary findings suggest that medical decision-making is an early feature of PSP.

Declaration of interest

None.

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References

[1] P. Appelbaum, T. Grisso, Assessing patients' capacities to consent to treatment, N. Engl. J. Med. 319 (1988) 1635–1638.
[2] K.L. Triebel, A. Gerstenecker, D.C. Marson, Neuropsychological assessment of decisional capacity in an aging society, National Academy of Neuropsychology Bulletin 28 (2014) 13–17.
[3] R.C. Martin, O.C. Okonkwo, J. Hill, H.R. Griffith, K. Triebel, A. Bartolucci, A.P. Nicholas, R.L. Watts, N. Stover, L.E. Harrell, D. Clark, D.C. Marson, Medical decision-making capacity in cognitively impaired Parkinson's disease patients without dementia, Movement Disorders, Official Journal Of The Movement Disorder Society 23 (13) (2008) 1867–1874.
[4] O.C. Okonkwo, H.R. Griffith, K. Belue, S. Lanza, E.Y. Zamrini, L.E. Harrell, et al., Cognitive models of medical decision-making capacity in patients with mild cognitive impairment, J. Int. Neuropsychol. Soc.: JINS 14 (2) (2008) 297–308.
[5] A. Gerstenecker, B.T. Mast, I. Litvan, Neuropsychiatric and cognitive disorders in other parkinsonian disorders, in: D. Aarsland, J. Cummings, D. Weintraub, K.R. Chaudhuri (Eds.), Neuropsychiatric and Cognitive Changes in Parkinson's Disease and Related Movement Disorders: Diagnosis and Management, Cambridge University Press, New York, NY, US, 2013, pp. 261–274.
[6] A. Gerstenecker, The neuropsychology (broadly conceived) of multiple system Atrophy, progressive supranuclear palsy, and corticobasal degeneration, Arch. Clin. Neuropsychol. : the official journal of the National Academy of Neuropsychologists 32 (7) (2017) 861–875.
[7] K. Duff, A. Gerstenecker, I. Litvan, Functional impairment in progressive supranuclear palsy, Neurology 80 (4) (2013) 380–384.
[8] A. Gerstenecker, K. Duff, B. Mast, I. Litvan, Behavioral abnormalities in progressive supranuclear palsy, Psychiatr. Res. 210 (3) (2013) 1205–1210.
[9] A. Gerstenecker, The Neuropsychology (Broadly Conceived) of Multiple System Atrophy, Progressive Supranuclear Palsy, and Corticobasal Degeneration, Archives of Clinical Neuropsychology, 2018.
[10] A. Gerstenecker, B. Mast, K. Duff, T.J. Ferman, I. Litvan, Executive dysfunction is the primary cognitive impairment in progressive supranuclear palsy, Arch. Clin. Neuropsychol. 28 (2) (2013) 104–113.
[11] A. Gerstenecker, E.D. Roberson, G.D. Schellenberg, D.G. Standaert, D.R. Sphrecher, B.M. Kluger, et al., Genetic Influences on Cognition in Progressive Supranuclear Palsy, (2017).
[12] I. Litvan, Y. Agid, D. Calne, G. Campbell, B. Dubois, R.C. Duvoisin, et al., Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report of the NINDS-SPSP international workshop, Neurology 47 (1) (1996) 1–9.
[13] G.U. Hoglinger, G. Respondek, M. Stamelou, Clinical diagnosis of progressive supranuclear palsy, The movement disorder society criteria 32 (6) (2017) 853–864.
[14] R.B. Postuma, D. Berg, M. Stern, W. Poewe, C.W. Olanow, W. Oertel, et al., MDS clinical diagnostic criteria for Parkinson's disease, Movement disorders : official journal of the Movement Disorder Society 30 (12) (2015) 1591–1601.
[15] I. Litvan, J.G. Goldman, A.I. Tröster, B.A. Schmand, D. Weintraub, R.C. Petersen, et al., Diagnostic criteria for mild cognitive impairment in Parkinson's disease: movement disorder society task force guidelines, Movement disorders : official journal of the Movement Disorder Society 27 (3) (2012) 349–356.
[16] G. McKhann, D. Drachman, M. Folstein, R. Katzman, D. Price, E.M. Stadlan, Clinical diagnosis of Alzheimer's disease: report of the NINCDS-ADRDA work group under the auspices of department of health and human services task force on Alzheimer's disease, Neurology 34 (7) (1984) 939–944.
[17] L.I. Golbe, P.A. Ohman-Strickland, A clinical rating scale for progressive supranuclear palsy, Brain : J. Neurol. 130 (Pt 6) (2007) 1552–1565.
[18] S. Fahn, R.L. Elton, Unified Parkinson's disease rating scale, in: S. Fahn, C.D. Marsden, D.B. Calne (Eds.), Recent Developments in Parkinson's Disease, Macmillan Health Care Information, Florham Park, NJ, 1987, pp. 153–164.
[19] D.C. Marson, K.K. Ingram, H.A. Cody, L.E. Harrell, Assessing the competency of patients with Alzheimer's disease under different legal standards. A prototype instrument, Arch. Neurol. 52 (10) (1995) 949–954.
[20] P.J. Jurica, C.L. Leitten, S. Mattis, Dementia Rating Scale - 2: Professional Manual, Psychological Assessment Resources, Lutz, FL, 2001.
[21] J. Brandt, R. Benedict, Hopkins Verbal Learning Test-revised: Professional Manual, PAR, Florida, 2001.
[22] S.M. Rao, N. Cognitive Function Study Group, A Manual for the Brief Repeatable Battery of Neuropsychological Tests in Multiple Sclerosis, National Multiple Sclerosis Society, New York, 1990.
[23] A. Gerstenecker, R. Martin, D.C. Marson, K. Bashir, K.L. Triebel, Introducing demographic corrections for the 10/36 spatial recall test, Int. J. Geriatr. Psychiatr. 31 (4) (2016) 406–411.
[24] R.M. Ruff, R.H. Light, S.B. Parker, H.S. Levin, Benton controlled oral word association test: reliability and updated norms, archives of clinical neuropsychology, The Official Journal Of The National Academy Of Neuropsychologists 11 (4) (1996) 329–338.
[25] J.H. Ricker, B.N. Axelrod, Analysis of an oral paradigm for the Trail making test, Assessment 1 (1) (1994) 47–51.
[26] B. Dubois, A. Slachevsky, I. Litvan, B. Pillon, The FAB: a frontal assessment battery at bedside, Neurology 55 (11) (2000) 1621–1626.
[27] A. Smith, Symbol Digit Modalities Test, Western Psychological Services, Los Angeles, CA, 1982.
[28] A. Gerstenecker, L. Niccolai, D. Marson, K.L. Triebel, Enhancing medical decision-making evaluations: introduction of normative data for the capacity to consent to treatment instrument, Assessment 23 (2) (2016) 232–239.
[29] O. Okonkwo, H.R. Griffith, K. Belue, S. Lanza, E.Y. Zamrini, L.E. Harrell, et al., Medical decision-making capacity in patients with mild cognitive impairment, Neurology 69 (15) (2007) 1528–1535.
[30] Research consent for cognitively impaired adults: recommendations for institutional review boards and investigators, Alzheimers Dis. Assoc. Disord. 18 (3) (2004) 171–175.