Progressive Supranuclear Palsy Rating Scale (PSPRS) with comments and instructions for administration

Golbe LI, Ohman-Strickland PA. A clinical disability rating scale for progressive supranuclear palsy. *Brain* 130:1552-1565, 2007.
Golbe LI. *A Clinician's Guide to Progressive Supranuclear Palsy*. Rutgers University Press, New Brunswick, NJ, 186 pages. December 2018.

Patient: _____ Date of birth: _____

Date: _____ Examiner: _____

Section, item title, definitions, item score box, section score box	Comments, instructions
History	
1. Withdrawal 0 None; baseline 1 Follows conversation in a group, may respond spontaneously, but rarely if ever initiates exchanges 2 Rarely or never follows conversation in a group ☐	• Ask caregiver • Use your own observation • Relative to baseline personality • Consider lack of conversation due to dementia or bradyphrenia as "withdrawal" • If patient never has opportunity to interact with people other than the caregiver, answer item by direct observation of patient's behavior in exam room.
2. Aggressiveness 0 No increase in aggressiveness 1 Increased, but not interfering with family interactions 2 Interfering with family interactions ☐	• Ask caregiver about frequent loss of temper, shouting • Relative to baseline personality • Ask if shouts or loses temper easily. • If resists caregiver's care but this does not risk patient's well-being, score a 1. If it does risk well-being, score a 2.
3. Dysphagia for solids 0 Normal; no difficulty with full range of food textures 1 Tough foods must be cut up into small pieces 2 Requires soft solid diet 3 Requires pureed or liquid diet 4 Tube feeding required for some or all feeding ☐	• Consider solid foods only • Ignore difficulty related to overloading mouth. • If the caregiver feels that solids must be cut smaller than typical for patient's size, score a 1. • If certain foods like bread crusts or leafy vegetables must be avoided, but meats are OK, score a 2. • Ignore the issue of patient's manual ability to cut food.

4. Using knife and fork, buttoning clothes, washing hands and face 0 Normal 1 Somewhat slow but no help required 2 Extremely slow; or occasional help needed 3 Considerable help needed but can do some things alone 4 Requires total assistance ☐	• Rate the worst of the 3 tasks. • If difficulty is related to downgaze, score as if it were purely motor. • If patient can do buttoning and washing and use a fork but not a knife, score a 3.
5. Falls 0 None in the past year 1 <1 per month; gait may otherwise be normal 2 1-4 per month 3 5-30 per month 4 > 30 per month (or chairbound) ☐	• Can ask patient or caregiver to estimate • Average frequency if attempted to walk unaided except for using walls and furniture. • Assume no access to walking aids. • Ignore near-falls.
6. Urinary incontinence 0 None or a few drops less than daily 1 A few drops staining clothes daily 2 Large amounts, but only when asleep; no pad required during day 3 Occasional large amounts in daytime; pad required 4 Consistent, requiring diaper or catheter awake and asleep ☐	• Ignore excuse of poor ambulation • If daytime pad used as precaution but no recent wetting, score a 3. • Define “a few drops” as staining underwear only. • If patient uses a pad “as a precaution” during the day, score a 3.
7. Sleep difficulty 0 Neither 1º nor 2º insomnia 1 Either 1º or 2º insomnia; averages ≥ 5 hours sleep nightly 2 Both 1º and 2º insomnia; averages ≥ 5 hours sleep nightly 3 Either 1º or 2º insomnia; averages < 5 hours sleep nightly 4 Both 1º and 2º insomnia; averages < 5 hours sleep nightly ☐	• 1º (primary) insomnia is difficulty falling asleep • 2º (secondary) insomnia is difficulty remaining asleep • Ignore trips to bathroom after which pt. returns to sleep easily. • This item may require asking 3 questions: falling asleep, staying asleep, average total duration • Ask caregiver about number of hours of sleep. OK to rely on subjective reports. • Ignore daytime sleep
History subtotal: ☐	

Mentation

Items 8-11 use this scale: 0 Clearly absent 1 Equivocal or minimal 2 Clearly present, but does not interfere with activities of daily living (ADL) 3 Interferes mildly with ADL 4 Interferes markedly with ADL	• Estimate the degree to which each deficit would interfere with performance of daily cognitive tasks.
8. Disorientation ☐	• Ask year, month, date, day, season, hospital, floor, city • Estimate or ask if problem dictates extra care from caregiver regarding ADLs.
9. Bradyphrenia ☐	• Observe delay for patient to answer easy questions • Estimate or ask if problem dictates extra care from caregiver re ADLs • If delayed responses prompt the caregiver to answer for the patient or limit your ability to interview patient, score at least a 3.
10. Emotional incontinence ☐	• If no emotional incontinence displayed, ask caregiver if patient laughs or cries when others usually do not • Count conversational interaction as an ADL • If there is a history of inappropriate laughing or crying but none at the time of the examination, score a 1 or 2, depending on its frequency. • If the conversation has to pause because of laughing or crying, score a 4.
11. Grasping/imitatative/utilizing behavior ☐	• Observe grasping of examiner or chair • Ask caregiver if grasping interferes with feeding, dressing • If none is displayed spontaneously (e.g., grabbing your coat or arm, or the wheelchair arm), ask patient to rest hands on thighs, palms up. Hold your hands 5-10 cm above his and say nothing. If he grabs them, rate a 3. • If he imitates your actions during the exam, rate a 2. • For an applause sign of at least 4 claps, rate a 1.
Mentation subtotal: ☐	

Bulbar

12. Dysarthria
0 None
1 Minimal; all or nearly all words easily comprehensible
2 Definite, moderate; most words comprehensible
3 Severe; may be fluent but most words incomprehensible
4 Mute; or a few poorly comprehensible words

☐

- Ignore palilalia and dysphonia
- "Comprehensible" means to examiner, not caregiver.
- If generally silent but can be coaxed to speak a few words, score a 4 no matter how clear those words may be.

13. Dysphagia
0 None
1 Single sips, or fluid pools in mouth or pharynx, but no choking/coughing
2 Occasionally coughs to clear fluid; no frank aspiration
3 Frequently coughs to clear fluid; may aspirate slightly; may expectorate frequently rather than swallow secretions
4 Requires artificial measures (oral suctioning, tracheostomy or feeding gastrostomy) to avoid aspiration

☐

- Give 30-50 cc of water in a cup, if safe.
- Do not use a straw. If patient or caregiver insists on a straw, score a 3.
- If patient finishes the cup without coughing, allow a few seconds more to observe.
- Do not give water if secretions are audible with breathing, if there is a history of frequent aspiration or if caregiver is apprehensive. In that case, score a 3.
- One cough scores a 2, multiple coughs scores a 3.

Bulbar subtotal: ☐

Ocular Motor

Items 14-16 use this scale:
0 Not slow or hypometric; 86-100% of normal amplitude
1 Slow or hypometric; 86-100% of normal amplitude
2 51-85% of normal amplitude
3 16-50% of normal amplitude
4 15% of normal amplitude or worse

14. Voluntary upward saccades

☐

15. Voluntary downward saccades

☐

16. Voluntary left and right saccades

☐

- Command the patient to "look up," "look down," "look right," "look left"
- For each, start from primary gaze, not the extreme opposite gaze
- Do not have patient follow a moving target or direct the gaze to a specific target However, use a target for primary gaze
- If improves with repetition, use the initial (i.e. worst) result
- For patients who fixate on your face, move away from the patient's primary gaze before testing
- If you can observe motion rather than just the starting and ending positions, it's "slow"
- If a corrective saccade is needed or if the motion is jerky, it's "hypometric"
- May hold lids to observe downward saccades
- Normal range of gaze is 50 degrees in each direction
- Ignore square-wave jerks and other intrusions
- If saccadic speed downward from the up to the primary position is worse than from primary to the down position, consider that "slow" for the purpose of supporting a score of 1

17. Eyelid dysfunction 0 None 1 Blink rate decreased (< 15/minute) but no other abnormality 2 Mild inhibition of opening or closing or mild blepharospasm; no visual disability 3 Moderate lid-opening inhibition or blepharospasm causing partial visual disability 4 Functional blindness or near-blindness because of involuntary eyelid closure ☐	• Observe patient in repose • Then have patient open/close lids quickly on command • Recruitment of frontalis muscle scores at least a 2. • Isolated difficulty closing lids on command scores at least a 2. • If patient keeps lids closed for several seconds at a time, score a 3.
Ocular Motor subtotal: ☐	

Limb motor

18. Limb rigidity 0 Absent 1 Slight or detectable only on activation 2 Definitely abnormal, but full range of motion possible 3 Only partial range of motion possible 4 Little or no passive motion possible ☐	• Score the worst of the four limbs • Count flexion contracture in advanced pts as dystonia, not rigidity • "Activation" means contralateral rapid alternating movement (fist opening or heel tapping)
19. Limb dystonia 0 Absent 1 Subtle or present only when activated by other movement 2 Obvious but not continuous 3 Continuous but not disabling 4 Continuous and disabling ☐	• Rate the worst of the 4 • Ignore neck and face • If no dystonia apparent on observation, have patient activate dystonia: • Hold hands out with eyes closed, palms down. Rate a finger extension or other sustained movement as 1. • Tap contralateral fingers or heel • Score the worst of the four limbs • Dystonia in PSP is usually asymmetric
20. Finger tapping 0 Normal (>14 taps/5 sec with maximal amplitude) 1 Impaired (6-14 taps/5 sec or moderate loss of amplitude 2 Barely able to perform (0-5 taps/5 sec or severe loss of amplitude) ☐	• If asymmetric, score worse side • Provide example, but stop and rate patient tapping alone • Have patient hold hand in range of his/her vision • If abnormal, encourage patient and score the best performance

21. Toe tapping 0 Normal (>14 taps/5 sec with maximal amplitude) 1 Impaired (6-14 taps/5 sec or moderate loss of amplitude 2 Barely able to perform (0-5 taps/5 sec or severe loss of amplitude) ☐	• If asymmetric, score worse side • Provide example and sit far enough from patient to allow his/her range of gaze to see your foot • If abnormal, encourage patient and score the best performance
22. Apraxia of hand movement 0 Absent 1 Present, not impairing most functions 2 Impairing most functions ☐	• If asymmetric, score worse side • Test for ideomotor apraxia • Two tasks with each hand (e.g., salute, throw ball, hitchhike, V-for-victory, wave good-bye) • Apraxic impairment should be evident on casual observation to score a 2. If it is evident only with testing, rate a 1.
23. Tremor in any part 0 Absent 1 Present, not impairing most functions 2 Impairing most functions ☐	• If tremor is not otherwise apparent, observe sustention and the finger-to-nose task • Rate the worse side • Upper extremities extended • Finger-to-nose with each hand
Limb Motor subtotal: ☐	
Midline/Gait	
24. Neck rigidity or dystonia 0 Absent 1 Slight or detectable only when activated by other movement 2 Definitely abnormal, but full range of motion possible 3 Only partial range of motion possible 4 Little or no passive motion possible ☐	• Rate the resistance to passive antero-posterior rotation. • You can ask patient to actively facilitate the passive movement • Ignore spontaneous posture (kyphosis, dystonic rotation, retrocollis) • If absent, attempt activation by opening/closing both fists • If there is no resistance over some of the range of movement but musculo-skeletal resistance thereafter, score a 0
25. Arising from chair 0 Normal 1 Slow but arises on first attempt 2 Requires more than one attempt, but arises without using hands 3 Requires use of hands 4 Unable to arise without assistance ☐	• Use an armless chair • Be ready to catch patient falling to front or back by putting yourself in front of patient with one hand ready to support neck from behind • If the patient must use hands, do not allow hands to contact the back of the chair – only the seat • If you don't have an armless chair, do not allow patient to push off the chair arms in arising – only the seat • Allow patient to reposition forward on the chair, but score at least a 2. Make an exception for short legs. • If cane/walker needed to arise, score a 4 • If can arise unassisted but falls forward, score a 4

26. Gait 0 Normal 1 Slightly wide-based or irregular or slight pulsion on turns 2 Must walk slowly or occasionally use walls or helper to avoid falling, especially on turns 3 Must use assistance all or almost all the time 4 Unable to walk, even with walker; may be able to transfer ☐	• Have patient walk down a hall if possible, not just across the exam room. • Include at least 1 pivot. • Tell the patient in advance where to pivot. • If patient can walk on a line with minimal difficulty but must use multiple careful steps to pivot, score a 2. • If must use walls/furniture to get across the exam room, score a 3. • If must use a walker or cane, score a 3.
27. Postural stability 0 Normal (shifts neither foot or one foot) 1 Must shift each foot at least once but recovers unaided 2 Shifts feet and must be caught by examiner 3 Unable to shift feet; must be caught, but does not require assistance to stand still 4 Tends to fall without a pull; requires assistance to stand still ☐	• Instruct patient to take a step with 1 foot and to try to keep the other foot planted • If can remain standing unassisted, explain test, pull backward by shoulders and be ready to catch him/her. Have a wall 1-2 meters behind you. • Pull should be hard enough to make a normal adult take 1 step back to retain balance. • Instruct patient to take 1 step with 1 foot and to try to keep the other foot planted.
28. Sitting down 0 Normal 1 Slightly stiff or awkward 2 Easily positions self before chair, but descent into chair is uncontrolled 3 Has difficulty finding chair behind him/her and descent is uncontrolled 4 Unable to test because of severe postural instability ☐	• Have patient approach chair from 3-4 steps away. • May touch seat but not arms or back of chair. • Keep your hands close to patient's back to prevent his missing the seat.
Gait/Midline subtotal: ☐	**PSPRS TOTAL:** ☐