

# **PSP-CDS**

# The Progressive Supranuclear Palsy Clinical Deficits Scale

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# The Progressive Supranuclear Palsy Clinical Deficits Scale

Patient Name/Subject ID		Assessment Date (mm/dd/yyyy)		Source of Information	
Functional Domain	0 = No Deficit	1 = Mild Deficit	2 = Moderate Deficit	3 = Severe Deficit	Score
Akinesia/rigidity	No akinesia or rigidity	Slow movements, but full range possible	Reduced range in active movements	Reduced range in passive movements	
Bradyphrenia	No bradyphrenia	Equivocal or mild, but not interfering with activities of daily living	Interfering moderately with activities of daily living	Interfering severely with activities of daily living	
Communication	No communicative dysfunction	Mild communicative dysfunction, but easily understood	Moderate communicative dysfunction, partly not understood	Severe communicative dysfunction, cannot be understood	
Dysphagia	No dysphagia	Mild dysphagia, but no dietary adaptations required	Moderate dysphagia, dietary adaptations required	Severe dysphagia, oral nutrition impossible	
Eye movements	No ocular motor dysfunction	Slow vertical saccades	Vertical supranuclear gaze palsy	Vertical & horizontal supranuclear gaze palsy	
Finger dexterity	No impairment in finger dexterity	Somewhat slow, but no help required when using knife and fork, buttoning clothes, washing hands and face	Extremely slow or occasional help required	Considerable help or total assistance needed	
Gait & balance	No postural instability	Postural instability, but unassisted gait possible	Gait possible with walking aid	Gait impossible	
Total Score					

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# **PSP-CDS User Instructions**

The Progressive Supranuclear Palsy Clinical Deficits Scale (PSP-CDS) provides a clinically meaningful measure to assess the functional impairment of patients with Progressive Supranuclear Palsy (PSP). The PSP-CDS is intended to monitor the patient's status in research and clinical practice.

The PSP-CDS covers seven functional domains (Akinesia/rigidity, Bradyphrenia, Communication, Dysphagia, Eye movements, Finger dexterity, Gait & balance) relevant to all clinical phenotypes of PSP and related diseases. All domains are rated for severity of deficits with scores ranging from 0 to 3, corresponding to no, mild, moderate, or severe deficits. Please rate all domains considering that increasing scores are intended to reflect an increasing impact on the patient's activities of daily living. The sum of individual domain scores provides a PSP-CDS Total Score ranging from 0 (no deficit in any domain) to 21 (severe deficit in all domains).

The information required for PSP-CDS rating is collected through a semi-structured interview with the patient and / or a reliable caregiver and a short structured clinical examination. The PSP-CDS User Instructions are specified in this document. The PSP-CDS table allows documenting the collected information.

# 1. Akinesia/rigidity

#### Examination

- **Evaluation of akinesia/bradykinesia** (as in MDS-UPDRS item 3.14): This combines all observations on slowness, hesitancy, and small amplitude and poverty of movement in general. This assessment is based on the examiner's global impression after observing for spontaneous gestures while sitting, rising, and walking.
- Evaluation of rigidity (as in MDS-UPDRS item 3.3): Rigidity is judged on slow passive movements of major joints with the patient in a relaxed position and the examiner manipulating the limbs and neck. First, test without an activation maneuver. If no rigidity is detected, use an activation maneuver such as tapping fingers, fist opening/closing, or heel tapping in a limb not being tested. Ask the patient to go as limp as possible as you test for rigidity.
- With varying scores in different body parts (e.g. limbs/trunk difference, asymmetry), rate the worst score.

#### Scores

- <u>O: No akinesia or rigidity</u>
  *Definition*: No akinesia or rigidity are detected during clinical examination in any body part.
- <u>1: Slow movements, but full range possible</u>
  *Definition*: Akinesia and / or rigidity are detected in at least one body part, but the corresponding movement can be performed in the full range.
- <u>2: Reduced range in active movements</u>

*Definition*: Akinesia and / or rigidity are detected in at least one body part, and the patient cannot voluntarily achieve the full range of movements in this body part, as would be expected for his / her age. However, the full range can be achieved passively by the examiner.

# • <u>3: Reduced range in passive movements</u>

*Definition:* Akinesia and / or rigidity are detected in at least one body part, and the patient cannot voluntarily achieve the full range of movements in this body part, as would be expected for his / her age. The full range also cannot be achieved passively by the examiner.

### 2. Bradyphrenia

### Examination

- Observe the patient during the entire consultation to judge slowing of thought processes, e.g. by increased latency to respond to simple questions. Rate symptoms that are persistent overtime rather than transient.
- Ask the patient and / or caregiver for an interference of slow thinking with activities of daily living (ADL) (e.g. solving of everyday problems, social interactions, hobbies, life at home, business and financial affairs).

# Scores

- <u>0: No bradyphrenia</u> *Definition*: No signs of slowed thinking.
- <u>1: Equivocal or mild, but not interfering with activities of daily living</u> *Definition*: Slowed thinking apparent to examiner, patient, or reliable caregiver. Activities of daily living are not affected.
- <u>2: Interfering moderately with activities of daily living</u>
  *Definition*: Slowed thinking apparent to examiner, patient, or reliable caregiver. Activities of daily
  living are affected, but the patient can still complete them without support from his / her caregiver.
- <u>3: Interfering severely with activities of daily living</u>
  <u>Definition</u>: Slowed thinking apparent to examiner, patient, or reliable caregiver. Activities of daily living are severely affected and the patient often requires support from his/her caregiver.

# 3. Communication

#### Examination

- Observe the patient during the entire consultation to judge oral communication abilities.
- Assess for presence of a <u>communication disorder</u> regardless of the underlying neurological cause (e.g. dysphonia, dysarthria, aphasia, apraxia of speech, apathy).
- In stressed or timid patients, rate communication at end of the consultation to allow the patient to calm down.
- In patients with language barrier, assess communication with the help of a reliable caregiver or translator.

#### Scores

- <u>0: No communicative dysfunction</u>
  <u>Definition</u>: Communicative dysfunction is not apparent during the consultation and not reported by the patient or a reliable caregiver.
- o <u>1: Mild communicative dysfunction, but easily understood</u>

*Definition*: Mild communicative dysfunction is apparent during the consultation, but the patient is easily understood.

- <u>2: Moderate communicative dysfunction, partly not understood</u>
  <u>Definition</u>: Moderate communicative dysfunction is apparent during the consultation. The patient can only be partly understood. Communication is effortful, but still efficient.
- <u>3: Severe communicative dysfunction, cannot be understood</u>
  *Definition*: Severe communicative dysfunction is apparent during the consultation. The patient cannot be understood. A few words or sentences may be understood, but effective communication is not possible.

# 4. Dysphagia

#### Examination

Observe the patient during the entire consultation to judge spontaneous dysphagia (e.g. frequency of coughing or harrumphing). Ask the patient and / or a reliable caregiver for the occurrence and frequency of aspirations and airway infections. Ask for the need of dietary adaptations (e.g. cutting solids into small pieces, pureed or thickened liquid diet) or parenteral feeding measures (e.g. tube feeding). A formal examination of dysphagia (e.g. by swallowing test or laryngoscopy) is not required for the purpose of this scale.

#### Scores

- <u>O: No dysphagia</u>
  <u>Definition</u>: No swallowing difficulties are being observed by the examiner, nor reported by the patient or a reliable caregiver.
- <u>1: Mild dysphagia, but no dietary adaptations required</u>
  *Definition:* Swallowing difficulties are being observed by the examiner, or reported by the patient or a reliable caregiver, but their severity is mild enough to require no dietary adaptations.
- <u>2: Moderate dysphagia, dietary adaptations required</u> *Definition:* Swallowing difficulties are being observed by the examiner, or reported by the patient or a reliable caregiver, severe enough to require dietary adaptations. The severity of moderate dysphagia may range from frequent coughing and requirement of small

dietary adaptations to the need of strained foods or thickened liquids. An increased frequency or severity of aspirations or airway infections caused by dysphagia, are also rated as 2, regardless of the patient's actual dietary adaptations, since that condition would in fact require adaptive measures.

• <u>3: Severe dysphagia, oral nutrition impossible</u> *Definition:* Daily oral nutrition is impossible. The patient requires parenteral feeding measures.

# 5. Eye movements

#### Examination

• **Range and velocity of gaze** should be assessed by command ("Look at the flicking finger") rather than by pursuit ("Follow my finger"), with the target >20 degrees from the position of primary gaze, in both the vertical (upward and downward) and horizontal (leftward and rightward) direction.

o In case of inconsistent results after repeated examinations, rate the worst result.

#### Scores

• <u>0: No ocular motor dysfunction</u>

*Definition*: No visible decrease in velocity and amplitude of vertical and horizontal saccadic eye movements. Ignore other eye movement dysfunctions (e.g. apraxia of eyelid opening, square wave jerks, nystagmus, blepharospasm).

• <u>1: Slow vertical saccades</u>

*Definition*: Decreased velocity of vertical greater than horizontal saccadic eye movements. Saccades are slow enough for the examiner to see the eye rotating, rather than just initial and final positions. A delay in saccade initiation is not considered as slowing. Findings are supported by slowed or absent fast components of vertical optokinetic nystagmus (i.e., only the slow following component may be retained). In case of slowing in any vertical direction (downward and / or upward), rate at least 1. Ignore horizontal slow saccades.

• <u>2: Vertical supranuclear gaze palsy</u>

*Definition*: A clear limitation of the range of voluntary gaze in the vertical plane in any direction (downward and / or upward), more than expected for age, which is overcome by activation with the vestibulo-ocular reflex.

• <u>3: Vertical & horizontal supranuclear gaze palsy</u>

*Definition*: A clear limitation of the range of voluntary gaze in both the vertical and in the horizontal plane, more than expected for age, which is overcome by activation with the vestibulo-ocular reflex; at late stages, the vestibulo-ocular reflex may be lost, or the maneuver prevented by nuchal rigidity.

# 6. Finger dexterity

#### Examination

 Observe the patient during the entire consultation to judge fine motor skills. Ask the patient and / or a reliable caregiver about the patient's independency in daily activities, particularly using knife and fork, buttoning clothes, washing hands and face (rate the worst). Ask about the difficulty in accomplishing those tasks, and the frequency and the extent to which the patient depends on external support for their accomplishment.

#### Scores

- O: No impairment in finger dexterity Definition: The patient is able to perform all above-mentioned tasks without slowness or impairment. Rate what you see and what the patient or a reliable caregiver report.
- <u>1: Somewhat slow, but no help required when using knife and fork, buttoning clothes, washing hands and face</u>

*Definition:* The patient is able to perform all above-mentioned tasks with some slowness or impairment. The patient is independent in those tasks and does not require outside help. Any outside assistance, regardless if provided by a relative, caregiver, or professional, is considered as help.

• <u>2: Extremely slow or occasional help required</u>

*Definition:* The patient is able to perform all above-mentioned tasks, but very slowly and with a lot of difficulty or may need occasional outside help.

<u>3: Considerable help or total assistance needed</u>
 *Definition:* The patient is unable to perform the majority of the above-mentioned tasks, being dependent on outside help most of the time.

# 7. Gait & balance

#### Examination

- Take the **patient's history** of the occurrence and frequency of unprovoked falls since onset of first PSP-related symptom. This information is important since some patients fall spontaneously despite normal gait assessment and pull-test. Note that the frequency of falls may decrease despite increasing disease severity due to preventive measures.
- **Gait assessment** (as in MDS-UPDRS item 3.10): Testing gait is best performed by having the patient walking away from and towards the examiner so that both right and left sides of the body can be easily observed simultaneously. The patient should try to walk 5 meters, then turn around and return to the examiner.
- **Pull-test** (as in MDS-UPDRS item 3.12): The test examines the response to a quick, forceful pull on the shoulders with the examiner standing behind the patient and the patient standing erect with eyes open and feet comfortably apart and parallel.

#### Scores

• <u>0: No postural instability</u>

*Definition*: No history of repeated, unprovoked falls, and no instability detected during gait assessment and in the pull-test (i.e., not more than 2 steps backwards).

- <u>1: Postural instability, but unassisted gait possible</u> *Definition*:
  - History of repeated, unprovoked falls since onset of first PSP-related symptom and / or
  - Signs of postural instability during gait assessment (swaying, stumbling, tendency to fall or seeking intermittent support while walking or turning) and / or
  - Signs of postural instability during the pull-test (more than 2 steps backwards or tendency to fall).
  - Despite these mild signs of postural instability, the patient is able to perform the gait assessment (as defined above) without assistance (no cane, walker, caregiver, or wall support).
- <u>2: Gait possible with walking aid</u>

*Definition*: Due to moderate postural instability, the patient is able to perform the gait assessment only with assistance (cane, walker, one caregiver, or wall support).

• <u>3: Gait impossible</u>

*Definition*: Due to severe postural instability, the patient cannot perform the gait assessment even with assistance (cane, walker, one caregiver, or wall support).