Published in final edited form as:

Curr Protoc Neurosci. 2009 October ; CHAPTER: Unit10.1. doi:10.1002/0471142301.ns1001s49.

Assessment of Parkinson Disease Manifestations

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Abstract

Parkinson disease (PD) is a progressive neurologic condition that causes motor and non-motor manifestations. Treatment provides symptomatic benefit but no current treatment has been proven to slow disease progression. Research studies of PD require a means of rating the severity of disease by measurement of motor manifestations, assessment of ability to perform daily functional activities, and symptomatic response to medication. The most common rating scales are the Unified Parkinson Disease Rating Scale (UPDRS), Hoehn and Yahr staging, and the Schwab and England rating of activities of daily living. Each of these rating scales are described, including detailed instructions on how to implement these ratings. Although these are the most widely applied rating scales of PD, there are still substantial limitations to these scales that must be considered when using them for research. Finally, some common applications of these scales are described.

Keywords

Parkinson disease; rating scales; activities of daily living; clinical trials

INTRODUCTION

Parkinson disease (PD) is a neurodegenerative disease that typically begins about age 60 and causes slowness of movement (bradykinesia), muscular stiffness (rigidity), tremor, poor postural stability, soft voice, shuffling gait, sudden cessation of movement called freezing, and a paucity of spontaneous movements (akinesia) (Fahn, 2003). Motor manifestations typically begin on one side of the body, only later affecting the other side as well (Rajput et al., 1991). Underlying degeneration of dopaminergic nigrostriatal neurons with subsequent striatal dopamine deficiency forms the basis for pharmacotherapy (McColl et al., 2002).

Oral levodopa or drugs that directly stimulate dopamine receptors provide symptomatic benefit that may be quite dramatic when the disease is relatively mild. As the disease progresses and treatment continues, treatment-related complications may develop, including short duration of response to individual doses of dopaminergic medication (Pahwa et al., 2006; Hershey et al., 2003). Patients are considered to be ON when medication is working and OFF when the benefit abates. The distinction between the ON and OFF phases can be quite dramatic, with an individual able to easily arise from a chair and literally run across the room, and then one minute later be unable to arise unassisted.

Other complications of treatment are involuntary, extraneous movements related to individual doses of medication. The most common types of these involuntary movements called dyskinesias are wiggly, twitchy, or sometimes smooth writhing movements that may occur either as medication is beginning to produce benefit, while it is working maximally, or as the benefit wears off. Another type of drug-induced involuntary movement is a more sustained type of muscle contraction typically with agonist and antagonist muscle co-contraction that may cause abnormal posturing of a part of the body. This type of movement, called dystonia, usually occurs when medication has worn OFF. However, it may also occur with the same

timing as dyskinesias. Other complications of dopaminergic drug therapy include development of nausea and vomiting, visual hallucinations, paranoid thinking, and exacerbation of orthostasis.

In addition to these obvious motor manifestations, there may be variable degrees of autonomic dysfunction such as orthostasis, constipation, urinary hesitancy with incomplete voiding, slowness of convergence (making it difficult to focus the eyes on nearby objects), heat intolerance, swallowing problems, and excessive drooling. Some patients also complain about vague, burning, achy types of pains called dysesthesias most commonly in the lower extremities (Chaudhuri et al., 2006). Important comorbid conditions include depression or dementia (Miyasaki et al., 2006).

There are several approaches used for clinical assessment of all of these manifestations of PD, and the most widely used are described here. Some scales assess motor manifestations or complications of treatment, and ratings may vary tremendously depending upon whether observations are recorded during the ON or OFF phase for those with such medication-induced fluctuations.

THE SCALES

Unified Parkinson Disease Rating Scale (UPDRS)

Multiple different scales for PD have been developed for quantification of motor manifestations (Webster, Columbia University Rating Scale, and Parkinson's disease Impairment Scale); disability (Schwab and England and Northwestern University Disability Scale); or both (UPDRS and New York University Scale) (Ramaker et al., 2002). Of these different scales, the UPDRS has gained the greatest acceptance as a tool for evaluation of interventions and as a clinical tool to follow patients (Martinez-Martin et al., 1994). However, there are important limitations to this scale (van Hilten et al., 1994; Goetz, 2004), and a new UPDRS is undergoing validation testing (Goetz et al., 2007).

The current UPDRS includes four subscales. Subscale 1 covers mentation, behavior, and mood. Subscale 2 rates activities of daily living. Subscale 3 is a clinician rating of the motor manifestations of PD. Subscale 4 covers complications of therapy. Data for subscales 1, 2, and 4 are elicited from patients and caregivers, whereas data for subscale 3 is examination-based. There are training tapes for the UPDRS subscales 2 (Goetz et al., 2003) and 3 (Goetz et al., 1995), and reviewing these can improve the reliability of the measures (Goetz and Stebbins, 2004). However, reliability of the other subscales depends on patient reporting in addition to examiner skills (Louis et al., 1996), but there is a training tape for the activities of daily living component subscale 2 (Goetz et al., 2003). The total UPDRS score and the UPDRS subscale scores are not interval scales, which means that there are not quantified, equal distances between values on these scales. For example, a score of 4 is greater than 2 but does not necessarily indicate twice the degree of severity. Each part of the rating is a rank order measure rather than a precise interval change. This must be considered when using these data for statistical analyses.

UPDRS Subscale 1: Mentation, Behavior, and Mood

The examiner asks the patient about each of the following areas of cognitive function or mood and the rater scores the answers from 0 to 4, with 4 representing the greatest level of dysfunction, based upon the responses of the patient or a caregiver. The sum of these scores for this subscale can range from 0 (normal) to 16.

1. Intellectual impairment (the list of possible responses can be read to the patient and examples provided as needed). Possible ratings of patient response:

0–none

1–mild consistent forgetfulness with partial recollection of events and no other problems

2–moderate memory loss, with disorientation and moderate difficulty handling complex problems

3–severe memory loss with disorientation with respect to time and often place; severe difficulty with complex problems

4–severe memory loss with orientation preserved only to person; unable to make judgments or solve problems, cannot be left home alone

2. Thought disorder (read the possible responses for the patient). Possible ratings of patient response:

0–none

1–vivid dreaming

2–benign hallucinations with insight preserved, that is, the patient is able to distinguish that the hallucinations are not real

3–occasional to frequent hallucinations or delusions with preserved insight; could interfere with activities of daily living

4–persistent hallucinations, delusions or florid psychosis, not able to care for self

3. Depression. Possible ratings of patient response:

0–not present

1–periods of sadness or guilt more than normal, but not sustained for more than 1 week

2–sustained depression greater than 1 week

3–sustained depression with vegetative symptoms

4–sustained depression with vegetative symptoms and with suicidal thoughts or intentions

4. Motivation and initiative. Possible ratings of patient response:

0–normal

1–less assertive than usual, more passive

2–loss of initiative or disinterest in elective activities (e.g., less interest in hobbies or social activities, tends to stay at home)

3–loss of initiative or disinterest in day-to-day activities (e.g., less attentive to personal hygiene, dressing)

4–withdrawn, complete loss of motivation (e.g., may sit in a chair most of the day with little initiative)

UPDRS Subscale 2: Activities of Daily Living

The examiner asks the patient to describe his or her function separately in the ON and OFF state. The responses for each of the 14 items on subscale 2 are therefore scored twice, once for ON and once for OFF. These ratings are done by the examiner based upon the responses of the patient or caregiver. The total score for subscale 2 ranges from 0 to 56.

1. Speech:
 - 0–normal
 - 1–mildly affected, with no difficulty being understood
 - 2–moderately affected, occasionally asked to repeat statements
 - 3–severely affected and frequently asked to repeat statements
 - 4–unintelligible most of the time
2. Salivation:
 - 0–normal
 - 1–slight but definite excess of saliva; may have nighttime drooling
 - 2–moderate excessive saliva; may have minimal daytime drooling
 - 3–marked excessive saliva; some daytime drooling
 - 4–marked drooling; requires constant use of tissue or handkerchief
3. Swallowing:
 - 0–normal
 - 1–rare choking
 - 2–occasional choking
 - 3–requires soft food
 - 4–requires nasogastric tube or gastroscopy tube for feeding
4. Handwriting:
 - 0–normal
 - 1–slightly slow or small
 - 2–moderately slow or small; all words are legible
 - 3–severely affected; not all words are legible
 - 4–the majority of the words are not legible
5. Cutting food and handling utensils:
 - 0–normal
 - 1–somewhat slow and clumsy but no help needed
 - 2–can cut most foods; although clumsy and slow; some help needed
 - 3–foods must be cut by someone, but can still feed self slowly
 - 4–needs to be fed
6. Dressing:
 - 0–normal
 - 1–somewhat slow, but no help needed
 - 2–occasional assistance needed with buttoning and putting arms into sleeves
 - 3–considerable help required, but can do some things alone

4—completely dependent upon assistance; unable to help

7. Hygiene:

0—normal

1—somewhat slow but no help needed

2—needs help to shower or bathe; very slow in hygienic care

3—requires assistance for washing, brushing teeth, combing hair, using the toilet

4—needs bladder catheter or other mechanical means

8. Turning in bed and adjusting bedclothes:

0—normal

1—somewhat slow and clumsy but no help needed

2—can turn alone or adjust sheets, but with great difficulty

3—can initiate attempt, but cannot turn or adjust sheets alone

4—cannot turn in bed or move a small amount to begin a turn or adjust bedclothes

9. Falling (unrelated to freezing):

0—none

1—rare falling

2—occasional falls; less than once daily

3—falls on average once daily

4—falls more than once a day

10. Freezing when walking:

0—none

1—rare freezing when walking; may have start hesitation

2—occasional freezing when walking

3—frequent freezing; occasional falls due to freezing

4—frequently falls due to freezing

11. Walking:

0—normal

1—mild difficulty; may not swing arms or tends to drag a leg

2—moderate difficulty but requires little or no assistance

3—severe disturbance of walking; requires assistance

4—cannot walk at all, even with assistance

12. Tremor in right arm:

0—absent

1—slight and infrequently present; not bothersome to patient

2—moderate, bothersome to patient

3–severe; interferes with many activities

4–marked; interferes with most activities

13. Tremor in left arm:

0–absent

1–slight and infrequently present; not bothersome to patient

2–moderate, bothersome to patient

3–severe; interferes with many activities

4–marked; interferes with most activities

14. Sensory complaints related to parkinsonism:

0–none

1–occasionally has numbness, tingling, or mild aching

2–frequently has numbness, tingling, or aching; not distressing

3–frequent painful sensations

4–excruciating pain

UPDRS Subscale 3: Motor Examination

Subscale 3 is an examiner rating of the motor manifestations of PD. This is the most commonly used subscale and has 14 different types of ratings, with many of these ratings done independently for the different limbs. Each of the ratings ranges from 0 to 4. The original UPDRS included only integers, but some use 0.5 increments; however, use of these 0.5 increments has not undergone clinimetric testing or validation. The total score for subscale 3 ranges from 0 to 108, the sum of scores from 27 observations. An accompanying video (Video 1) shows how the first 20 items are rated, and a second video (Video 2) illustrates the last 7 items on this motor subscale.

Tremor at rest, items 1 to 5—Tremor at rest is rated in the face and each extremity separately (5 ratings). The typical facial resting tremor appears in the chin, jaw, or lips, and is best observed with the patient seated on a chair. To assess tremor in the upper extremities, ask the patient to rest the forearms comfortably on the thighs. Resting tremor most commonly appears as a flexion-extension movement of the wrist/hand, a pronation-supination movement of the forearm, or a pill-rolling movement of the thumb and index finger. For some people, this tremor may only be apparent while walking. Tremor in the legs may be visible either with the legs at rest while sitting or when lying supine. Ratings are then done for the face and each upper extremity separately using the following scoring:

0–none

1–slight or infrequently present

2–mild in amplitude or persistent, or moderate in amplitude and intermittent

3–moderate in amplitude and present most of the time

4–marked in amplitude and present most of the time

Action tremor, items 6 and 7—Action or postural tremor of the hands is assessed with the upper extremities under three conditions. First the upper extremities are fully extended straight ahead at shoulder height with the index fingers of either hand pointing at each other, held

closely but not quite touching. The next position for assessment is with the elbows fully flexed and the arm elevated at the patient's sides (like "chicken wings") and the index fingers held close to either side of the nose, without touching the nose. Finally, the patient should be instructed to touch the index finger to the nose and then fully extend the limb to use that same index finger to touch the examiner's finger. This motion should be repeated several times with each limb. After all of these maneuvers, a single rating is done for each limb for action or postural tremor:

- 0—none
- 1—slight, present with action
- 2—moderate in amplitude, present with action
- 3—moderate in amplitude, present with posture holding as well as with action
- 4—marked in amplitude, sufficiently severe to interfere with feeding

Rigidity, items 8 to 12—Rigidity refers to increased resistance to passive range of motion and is rated for the neck and all four extremities separately (5 ratings). Rigidity feels like bending a lead pipe; occasionally there is a ratchetiness called cogwheeling that also may be felt. Rigidity is tested in the limbs by supporting the joint with one hand; the examiner then uses the other hand to move the patient's limb through a full range of motion. In the upper extremities, this is tested at the wrist, elbow, and shoulders with the greatest amount of rigidity detected used as the rating for that limb. Rigidity in the lower extremities is usually tested by fully extending and flexing the knee with the patient either sitting or lying supine. Rigidity at the neck is tested in the seated patient by the examiner placing one hand on the forehead and the other hand on the occipital part of the head. The head is then gently fully extended and flexed with the degree of resistance assessed. If no rigidity is detected at one of these sites, then it should be checked again while either having the patient make large circles in the air with a limb on the opposite side of the body or while tapping either foot on the floor. These additional movements increase rigidity. Separate ratings for each of the limbs and the neck are done using the following scoring:

- 0—none
- 1—slight or detectable only when activated by other movements
- 2—mild to moderate
- 3—marked but full range of motion easily achieved
- 4—marked and full range of motion achieved with difficulty

Bradykinesia, items 13 to 14—Bradykinesia, or slowness of repetitive movements, is rated using three different methods in the upper extremities, rating the left and right separately (and testing each area separately). The first test is finger tapping. The patient is seated and asked to tap the thumb and index finger, making the movements as wide and fast as possible. This is done for at least 15 sec with each hand. The speed of movement and the amplitude are included in a single rating for each hand.

- 0—normal (>15/5 sec)
- 1—mild slowing and/or reduction of amplitude (11 to 14/5 sec)
- 2—moderate in amplitude; definite early fatiguing; may have occasional arrests of movement (7 to 10/5 sec)

3—severely impaired; frequent hesitation in initiating movements or arrests in ongoing movement (3 to 6/5 sec)

4—can barely perform the task (0 to 2/5 sec)

Bradykinesia, items 15 to 16—Bradykinesia is also rated with the arms in the same position as for hand rotation, but this time having the patient open and close the hand as quickly as possible, also with the largest excursion possible. Ratings for each hand should be done.

0—normal (>15/5 sec)

1—mild slowing and/or reduction of amplitude (11 to 14/5 sec)

2—moderate in amplitude; definite early fatiguing; may have occasional arrests of movement (7 to 10/5 sec)

3—severely impaired; frequent hesitation in initiating movements or arrests in ongoing movement (3 to 6/5 sec)

4—can barely perform the task (0 to 2/5 sec)

Bradykinesia, items 17 to 18—Bradykinesia is also rated for each upper extremity by having the seated patient raise the elbow to the level of the mid-chest, flex it to 90° with the hand pointing up, and then rotate the hand and forearm as rapidly as possibly with the greatest excursion possible. This motion should also continue for 15 sec. This is rated in the same fashion as finger tapping for each side.

0—normal (>15/5 sec)

1—mild slowing and/or reduction of amplitude (11 to 14/5 sec)

2—moderate in amplitude; definite early fatiguing; may have occasional arrests of movement (7 to 10/5 sec)

3—severely impaired; frequent hesitation in initiating movements or arrests in ongoing movement (3 to 6/5 sec)

4—can barely perform the task (0 to 2/5 sec)

Bradykinesia, items 19 to 20—Bradykinesia is also tested in the feet with the patient seated and having both feet flat on the floor. Then, each foot is tested separately by having the patient raise the entire foot and lower it back to the floor with as wide excursion as possible and as fast as possible. Some have modified this to have the patient keep the heel on the floor and then repetitively elevate the front of the foot by dorsiflexing the ankle, and lastly return the foot flat on to the floor—again using the largest excursion possible and as fast as possible. Each limb is then scored.

0—normal (>15/5 sec)

1—mild slowing and/or reduction of amplitude (11 to 14/5 sec)

2—moderate in amplitude; definite early fatiguing; may have occasional arrests of movement (7 to 10/5 sec)

3—severely impaired; frequent hesitation in initiating movements or arrests in ongoing movement (3 to 6/5 sec)

4—can barely perform the task (0 to 2/5 sec)

Speech, item 21—Speech typically becomes soft and indistinct in PD. This is rated by listening to the patient speak, but there are no standard sentences that have been used for this rating. Overall, consistency of this rating is improved by identifying a standard short paragraph for the patient to read and using this same paragraph on subsequent ratings. Speech is scored with a single rating.

0—normal

1 – slight loss of expression, diction, or volume

2—monotone, slurred but understandable, moderately impaired

3—marked impairment, difficult to understand

4—unintelligible

Facial expression, item 22—Facial expression is rated by observing the spontaneous expressive movements of the face during the entire evaluation. People with PD tend to develop reduced spontaneous facial expression and a mask-like face.

0—normal

1—minimal hypomimia (reduced facial movement); may be a normal “poker face”

2—slight, but definitely abnormal reduction of facial expression

3—moderate hypomimia; lips parted some of the time

4—masked or severe hypomimia with severe or complete loss of facial expression; lips parted ¼ inch or more

Arising from a chair, item 23—Ability to arise from a chair is rated by having the patient first cross the arms on the chest and then try to arise from a firm chair that has solid arm rests, but without using the arms to assist, if possible. Degree of disability is rated.

0—normal

1—slow or may need more than one attempt

2—pushes self up from arms of seat

3—may fall back and may have to try more than once, but able to do this independently

4—unable to arise without help

Posture, item 24—Posture tends to become increasingly flexed at the neck, shoulders, elbows, wrists, hips, and knees as PD progresses. A single rating for posture as a whole is done.

0—normal

1—not quite erect, slightly stooped; could be normal for an elderly person

2—moderately stooped; definitely abnormal; can be slightly leaning to one side

3—severely stooped with kyphosis; can be moderately leaning to one side

4—marked flexion with extreme abnormality of posture

Gait, item 25—As PD progresses, gait is characterized by smaller, slower steps with a shuffling character. Armswing may be reduced while walking. Some patients tend to shuffle faster and faster while leaning forward as walking continues. This is called propulsion. Others

tend to fall and take many steps backwards, so called retropulsion. Festination refers to shuffling, hesitant steps. All of these features are considered in a single rating of gait.

0—normal

1—walks slowly; may shuffle with short steps but no festination or propulsion

2—walks with difficulty but requires little or no assistance; may have some festination, short steps, or propulsion

3—severe disturbance of gait; requires some assistance

4—cannot walk at all, even with assistance

Postural stability, item 26—Postural stability is assessed with the pull test. People with PD tend to have increasing difficulty maintaining balance, particularly with a backwards perturbation. The pull test is done to determine an individual's ability to maintain upright posture with a sudden backwards pull. The test is done as follows. First the patient is asked to stand with the feet shoulder width apart and eyes open. The examiner stands behind the subject in a position that permits the examiner to brace him or herself in case the patient falls backwards during the testing procedure. An examiner can stand with the back close to a wall if there is concern about not being able to catch the patient falling backwards. Once in position, the examiner instructs the patient to resist a backwards pull, but, if needed, step backward to maintain balance. The examiner then gives a small practice pull on the patient's shoulder. After that, the patient is warned that the next pull will be much harder and then the examiner gives a sudden strong backwards pull on both shoulders. If the patient cannot maintain balance while standing before any pull, then the pull part of the test is not necessary. The pull test is given a single rating.

0—normal (takes two or fewer steps backward to maintain balance)

1—takes more than two backwards but stops self from falling

2—would fall if not caught by the examiner

3—very unstable; tends to lose balance spontaneously

4—unable to stand without assistance

Body bradykinesia, item 27—People with PD also tend to have fewer spontaneous movements (so-called akinesia) and generally slow movements. These are rated for the body as a whole with a single rating (note that akinesia is also rated by facial expression).

0—normal

1—minimal slowness giving movements a deliberate character, possibly reduced amplitude

2—moderate degree of slowness and poverty of movement that is definitely abnormal; alternatively some reduced amplitude

3—moderate slowness, poverty, or small amplitude of movement

4—marked slowness, poverty, or small amplitude of movement

UPDRS Subscale 4: Complications of Therapy

Subscale 4 includes 11 questions that relate to the observations of the patient or caregiver during the preceding 1 week. The first three questions and question 8 are rated from 0 to 4, but the remaining questions are simple no/yes questions. The score on this subscale ranges from 0 to 23. Note, however, that this is a relatively crude measure of dyskinesias and is not based upon direct examiner observation. One important limitation of this approach is that patients may not

be aware they are having dyskinesias. In these situations, it will be helpful to interview a caregiver or other observer to obtain collateral information. In fact, it may be necessary to demonstrate those movements to this collateral source.

Dyskinesias

Duration of dyskinesias, item 1: Record the proportion of the waking day during which dyskinesias are present:

- 0—none
- 1—1% to 25% of the day
- 2—26% to 50% of the day
- 3—51% to 75% of the day
- 4—76% to 100% of the day

Disability, item 2: Rate disability due to dyskinesias:

- 0—not disabling
- 1—mildly disabling
- 2—moderately disabling
- 3—severely disabling
- 4—completely disabling

Pain, item 3: Rate pain that the patient associates with dyskinesias:

- 0—not painful
- 1—slightly painful
- 2—moderately painful
- 3—severely painful
- 4—markedly painful

Early morning dystonia, item 4: Record presence of early morning dystonia (usually cramping in the legs or twisting of ankle that occurs when first awakening in the morning):

- 0—no
- 1—yes

Clinical fluctuations

Predictability, item 5: Are any OFF periods predictable as to timing after a dose of medication?

- 0—no
- 1—yes

Unpredictable OFF, item 6: Are any OFF periods unpredictable as to timing after a dose of medication?

- 0—no

1–yes

Sudden OFF, item 7: Do any OFF periods come suddenly (e.g., within a few seconds)?

0–no

1–yes

Time OFF, item 8: What proportion of the waking day is the patient OFF, on average?

0–none

1–1% to 25% of the day

2–26% to 50% of the day

3–51% to 75% of the day

4–76% to 100% of the day

Other complications

GI complaints, item 9: Does the patient have anorexia, nausea or vomiting?

0–no

1–yes

Sleep disturbances, item 10: Does the patient have any sleep disturbances (e.g., insomnia or hypersomnolence (i.e., sleeping through the night or excessive daytime sleepiness)?

0–no

1–yes

Orthostasis, item 11: Does the patient have symptomatic orthostasis (i.e., a sense of light-headedness or actual fainting due to a drop in blood pressure associated with standing)?

0–no

1–yes

Hoehn and Yahr Rating Scale

The Hoehn and Yahr staging is probably the most widely known evaluation of people with PD and was first described in 1967 (Hoehn and Yahr, 1967). It is really a simple staging from 0 to 5 of the motor manifestations of PD, intended to reflect the degree of progression, and combines features of motor impairment and disability. However, the scale is not linear and may not even be rank order, with some people having greater disability with stage 2 (with substantial bradykinesia but good stability on the pull test) compared to some that have been ranked as stage 3 (that fall on the pull test but have relatively mild bradykinesia and rigidity). The rating was subsequently modified to include two half scores, as noted below, but no clinimetric testing of this modification has been done (Goetz et al., 2004). Overall, the Hoehn and Yahr staging is best used for a description of subject groups, and any analysis should use nonparametric statistics. This rating is based upon examination of the patient.

0–no signs of disease

1–unilateral disease (on one side)

1.5–unilateral disease plus axial involvement

- 2–bilateral disease, without impairment of balance
- 2.5–bilateral disease, with recovery on the pull test
- 3–mild to moderate bilateral disease; needs assistance to prevent falling on pull test; physically independent
- 4–severe disability, but still able to walk or stand unassisted
- 5–wheelchair-bound or bedridden unless aided

The major strength of this scale is that it is well known and the tests are easily performed. However, this is a categorical scale and may not always be rank order, i.e., someone with stage 2 who is very slow with much tremor but has intact postural reflexes may be more impaired for activities of daily living than someone rated as stage 3 who is not nearly as slow or tremulous but falls on the pull test.

Schwab and England ADL Scale

The Schwab and England Scale is an “activities of daily living” (ADL) scale frequently used to provide a single estimate of the patient’s ability to function. The rating is done by the examiner interviewing the patient and, frequently, a collateral source, such as a spouse. This rating varies from 0 to 100% using 5% increments.

- 100%–completely independent; able to do all chores without slowness, difficulty, or impairment; essentially normal; unaware of any difficulty
- 90%–completely independent and able to do all chores with some degree of slowness, difficulty, or impairment; some activities might take twice as long; beginning to be aware of difficulty
- 80%–completely independent in most chores; some activities take twice as long; conscious of difficulty and slowness
- 70%–not completely independent; more difficulty with some chores; some tasks now take three to four times as long; must spend a large part of the day with chores
- 60%–some dependency; can do most chores, but exceedingly slowly and with much effort; some tasks cannot be done; common errors
- 50%–more dependent; needs help with about half of activities; slower and experiencing difficulty with all tasks
- 40%–very dependent, but still able to assist with all chores; however, few can be done independently
- 30%–all tasks require much effort; a few chores can be done alone or at least started alone; much assistance needed
- 20%–no tasks done independently; patient can provide slight help with some chores; but requires substantial assistance for all activities
- 10%–totally dependent and requires assistance with all activities of daily living
- 0%–vegetative functions with loss of control of swallowing, bladder, and bowel functions; bedridden

APPLICATIONS OF THE RATING SCALES

The need for a metric to assess therapeutic interventions was one of the key driving forces behind development of rating scales. In general, rating scales can be divided into those that reflect functional fluctuations (like the UPDRS subscale 3 and Hoehn and Yahr scales) and

those that use patient or caregiver reports to provide a general level of function (like Schwab and England). Since symptomatic effects of drugs can cause marked changes in ratings, the use of such ratings to indicate disease progression must attempt to control for these transient, treatment-related fluctuations. This applies to any clinical trial of an intervention, as well as utilization of ratings as a clinical tool to assess progression for clinical care. This section describes some of the strategies needed to address these issues.

Assessment of Symptomatic Effects

Ratings such as the UPDRS, either in its entirety or with motor subscale 3 alone, have been used to assess symptomatic benefit from therapeutic interventions. A critical factor in longitudinal studies is to try to control the variables in the temporal responses to individual doses of medication. Ideally, repeated ratings should be done at the same time of day, at the same time after the last dose of medication, and by the same rater. Other factors also may influence these ratings, such as diet, fatigue, or other potential stresses that may alter PD manifestations.

Monitoring Disease Progression

Attempts have been made to use UPDRS ratings to monitor disease progression or to evaluate the efficacy of a potentially disease-modifying intervention. In these cases, it is important to minimize the symptomatic effects of either the medication under investigation or of concomitant drugs. Much of the symptomatic benefit of levodopa diminishes over a day, and many studies choose to rate patients in the morning after an overnight abstinence from other PD medications (Nutt and Woodward, 1986; Chan et al., 2004). This approach has not only been applied to multiple drug trials, but also to assess effects of fetal cell transplantation to minimize effects of concomitant medication (Langston et al., 1992). However, this approach does not completely eliminate symptomatic effects of levodopa, which may gradually diminish over the course of a week (Chan et al., 2004). However, such prolonged washout may be impractical, depending upon the severity of the PD in patients in the study (Fahn et al., 2004).

Use of Rating Scales for Screening for Parkinsonism

Although the UPDRS was not developed for diagnostic use, it has been applied for mass screenings for parkinsonism (Racette et al., 2005). Note the use of the term “parkinsonism” rather than PD. Parkinsonism refers to the clinical manifestations, but makes no specific inference regarding the pathophysiology or etiology of the condition. Thus, these types of mass screenings attempt to identify the more general condition “parkinsonism” that may include drug-induced parkinsonism, multisystems atrophy, or progressive supranuclear palsy, as well as Parkinson disease. Furthermore, ratings on the UPDRS can reflect nonparkinsonian conditions. A hemiparesis due to a stroke may enhance or produce bradykinesia in a limb that is not distinguished from parkinsonism on the rating scale. Nevertheless, there have been several attempts to use modified forms of UPDRS subscale 3 as a screening for parkinsonism (Racette et al., 2006). In fact, mass screenings can include blinded videotape-based ratings that exclude assessment of rigidity, since that must be done in person. The key feature is to determine what value of UPDRS subscale 3 indicates parkinsonism. A score of nine on subscale 3 has been demonstrated to have 100% sensitivity and 81% specificity in identification of parkinsonism in a large video-based screening of more than 2000 people with welding exposure, an environmental factor under investigation for its potential relationship to the development of parkinsonism.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

This work was supported by NIH grants RR024992, NS050425, and NS041509 and the Greater St. Louis Chapter of the American Parkinson Disease Association (APDA), the APDA Center for Advanced PD Research at Washington University, the Barnes-Jewish Hospital Foundation (The Jack Buck Fund and the Elliot H. Stein Family Fund), the Missouri Chapter of the Dystonia Medical Research Foundation, and the Sam and Barbara Murphy Fund.

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