Movement Disorders
Vol. 18, No. 3, 2003, pp. 303–312
© 2002 Movement Disorder Society

Rating Scales for Dystonia: A Multicenter Assessment

Cynthia L. Comella, MD,* Sue Leurgans, PhD, Joanne Wuu, ScM, Glenn T. Stebbins, PhD, Teresa Chmura, BA and The Dystonia Study Group†

Rush-Presbyterian–St. Lukes Medical Center, Chicago, Illinois, USA

Abstract: The evaluation of dystonia requires a reliable rating scale. The widely used Fahn-Marsden Scale (F-M) has not been sufficiently tested across multiple centers and investigators. The Dystonia Study Group developed the Unified Dystonia Rating Scale (UDRS) and a Global Dystonia Rating Scale (GDS) to serve as instruments to assess dystonia severity. In this study, 25 dystonia experts evaluated the UDRS, F-M, and GDS for internal consistency and reliability. One hundred dystonia patients were videotaped using a standardized videotape protocol. Each examiner rated 20 patients using the UDRS, F-M, and GDS in random order. The examiner then assessed each scale for ease of use. Statistical analysis used Cronbach's α, intraclass correlation coefficients (ICC), generalized weighted κ statistic, and Kendall's coefficient of concordance. The UDRS, F-M, and GDS showed excellent internal consistency (Cronbach's α 0.89–0.93) and good to excellent correlation among the raters (ICC range from 0.71–0.78). Inter-rater agreement was fair to excellent (Kendall's 0.54–0.87; κ 0.37–0.91) being lowest for eyes, jaw, face, and larynx. The modifying ratings (Duration in the UDRS and Provoking Factor in the F-M) showed less agreement than the motor severity ratings. Among scales, the total scores correlated (Pearson's *r*, 0.977–0.983). Overall, 74% of raters found the GDS the easiest to apply. The GDS with its simplicity and ease of application may be the most useful dystonia rating scale. © 2002 Movement Disorder Society

Key words: dystonia; rating scale; movement disorder; outcome assessment

Dystonia is defined as a syndrome consisting of involuntary movements characterized by twisting or sustained movements.[1] It is a dynamic condition that often changes in severity depending on the posture assumed and activity of the involved body area. The changing nature of dystonia makes the development of rating scales with acceptable clinometric properties problematic.[2] The Fahn-Marsden rating scale (F-M) was the first dystonia scale evaluated for its clinometric properties. In a study using 10 dystonia patients and 4 raters, the reliability, inter-rater agreement, and concurrent validity of the F-M were demonstrated for the total score without reporting the level of agreement for ratings of the different body regions.[3] Although promising, the F-M scale was never assessed further as a multi center instrument that could be used by many investigators. Furthermore, the small number of dystonia patients included may not have represented the full spectrum of dystonia severity that would be encountered in a multicenter study. Some of the limitations in the F-M include the variable definition of body areas, and a weighting factor of 0.5 that halves the contribution of dystonia in eyes, mouth and neck to the total score. Recognizing these potential limitations, the Dystonia Study Group (DSG) designed a new rating scale, the Unified Dystonia Rating Scale (UDRS) that addressed these issues. A DSG consensus conference in 1997 produced the UDRS and a standardized protocol for videotaping dystonia patient. The global dystonia rating scale (GDS) was also created.

The UDRS was designed to include a more detailed assessment of individual body areas, including separate ratings for proximal and distal limbs, and elimination of the subjective patient rating for speech and swallowing included in the F-M. In addition, a duration rating was developed that paralleled a duration factor previously

† See Appendix for a list of Study Participants.

*Corresondence to: Cynthia L. Comella, Department of Neurological Sciences, Rush Presbyterial-St.Luke's Medical Center, Suite 755, 1725 West Harrison, Chicago, IL 60612. E-mail: ccomella@rush.edu

Received 5 July 2002; Revised 29 August 2002; Accepted 29 August 2002

validated within the Toronto Western Spasmodic Torticollis Scale (TWSTRS).[4–6] In contrast to the F-M, the UDRS has no weighting factors for any body region. The GDS is a global scale applied to individual body regions.

In this DSG-initiated study, the specific aims were to evaluate the internal consistency, inter-rater reliability and clinical applicability of the F-M, UDRS, and GDS across multiple sites in a large number of dystonia patients encompassing the full spectrum of dystonia severity.

PATIENTS AND METHODS

Patients

Patients were included in the study if they had primary dystonia and were followed in the outpatient Movement Disorders clinic offices at Rush-Presbyterian-St. Luke's Medical Center. Informed consent, as approved by the Institutional Review Board at Rush Presbyterian St. Luke's Medical Center, was obtained from all participants. The diagnosis of primary dystonia was based on presence of dystonia and absence of additional neurological signs or causes for dystonia. In particular, attention was paid to recruiting patients with generalized dystonia so that each master tape could have adequate representation of types and range of dystonia.

Investigators

The 25 rating investigators from 20 institutions are listed in Appendix 1. The Principal Investigator (CLC) was not included as a rating investigator, and carried out the initial screening of the videotapes of all patients. There were no investigators from the recruiting institution to prevent any rater from having previous knowledge of the patients included for rating. Each investigator was a specialist in Movement Disorders with expertise in the evaluation of dystonia.

Videotaping Protocol and Development of Rating Tapes

There were 103 patients videotaped using a standard videotape protocol that incorporates and expands the videotape protocol included with the F-M (see Appendix 2).[3] The videotape protocol includes examination of each body region at rest and during activation procedures. Patients were videotaped in a uniform manner. All the videotapes were evaluated by the PI (CLC) who rated each of 10 body areas for severity of dystonia using a 0 to 10 scale, with 0 defined as no dystonia and 10 as severe dystonia. These scores were used to allocate patients to a severity level and then randomly allocate patients such that each master evaluation tape included a range of dystonia severity. Each investigator rated two master evaluation tapes with 10 patients included on each tape, or a total of 20 patients. No pair of investigators rated the same two tapes. A statistician (SL) used computer-generated random numbers to allocate pairs of tapes to the raters. The rating investigator viewed the evaluation videotapes a total of three times using the UDRS for rating during one viewing, the GDS during another viewing and the F-M during another viewing. The order of scale application was randomized.

Rating Scales

The UDRS includes ratings for 14 body areas including eyes and upper face, lower face, jaw and tongue, larynx, neck, trunk, shoulder/proximal arm (right and left), distal arm/hand (right and left), proximal leg (right and left), and distal leg/foot (right and left) (Appendix 3). For each of the 14 body areas assessed, the UDRS has a severity and a duration rating. The severity rating is specific for each body region assessed and ranges from 0 (no dystonia) to 4 (extreme dystonia). The duration rating is modified from the duration factor of the TWSTRS, and ranges from 0 to 4. The Duration rating assesses whether dystonia occurs at rest or with action, and whether it is predominantly at maximal or sub maximal intensity. The total score for the UDRS is the sum of the severity and duration factors. The maximal total score of the UDRS is 112.

The F-M rating scale (see Appendix 4) evaluates dystonia in nine body areas, including eyes, mouth, speech and swallowing, neck, trunk, and right and left arm and leg. The arms and legs are given one rating each, without distinguishing proximal and distal elements. For each of the nine body regions, severity ratings range from 0 (no dystonia) to 4 (severe dystonia). The provoking factor rating assesses the situation under which the dystonia occurs and ranges from 0 (no dystonia) to 4 (dystonia at rest). The score for the eyes, mouth, and neck are each multiplied by 0.5 before being entered into the calculation of the total score. The total score of the F-M is the sum of the products of the provoking, severity and weighting factors. The maximal total score on the F-M is 120.[2]

The GDS rates dystonia severity in the 14 body areas already described for the UDRS (see Appendix 5). The GDS is a Likert type scale with ratings from 0 to 10 (0 is no dystonia, 1 minimal, 5 moderate and 10 severe dystonia) (Appendix 4). There are no modifying ratings or weighting factors in the GDS. The total score is the sum of the scores for all the body areas. The maximal total score of the GDS is 140.

After ratings were completed using all three scales, each investigator completed a standard questionnaire for each scale that assessed the investigator's opinion of ease of application, usefulness in an office setting and usefulness in multicenter trials.

Statistical Analysis

Analyses were done using the statistical software *SAS v. 6.12*, *STATA v. 6.0*, or *SAS% MAGREE* macro where appropriate (Stata Corp., College Station, TX; SAS Inc., Cary, NC). The total score by rater for each patient was calculated for each scale. The ratings were averaged across the five raters for each patient. Summary statistics of the overall scores are presented as mean ± SD ratings and range, Pearson's correlation were used for pair-wise comparison of the total scores of the three scales.

The internal consistency of each scale was assessed by Cronbach's α. Overall inter-rater agreement was assessed using intraclass correlation coefficient (ICC). The ICC was first computed for each tape (containing 10 distinct subjects) as rated by the five raters. The overall ICC was calculated by averaging across the 10 tapes.

Inter-rater agreement for body regions was analyzed in two ways: Kendall's coefficient of concordance and generalized weighted κ. To show the agreement for comparable body regions, the UDRS and GDS ratings for 2 areas (proximal and distal limbs; and jaw, lower face and mouth) were collapsed and the more severe score used.

The Kendall's coefficient of concordance provides a measure of the consistency among raters in the rankings of dystonia severity. Kendall's coefficient of concordance for each body region was computed for each tape, then averaged across tapes.

The generalized weighted κ statistic provides a measure of agreement in absolute ratings among more than two raters and on a scale with more than two rating categories. In this study, Kappa was computed using four rating groupings to allow stable calculations: GDS 0–1, 2–3, 4–6, 7–10; UDRS 0, 1, 2, 3–4; F-M 0, 1, 2, 3–4. Kappa values exceeding 0.75 are usually considered excellent agreement, values between 0.4 and 0.75 fair to good agreement, and values below 0.4 poor agreement.[10]

For both the Kendall's coefficient of concordance and the generalize weighted κ, an outcome of 0 indicates no agreement beyond chance, and 1 indicates perfect agreement.[7] Reliability and inter-rater agreement were analyzed separately for severity and the modifying factors (UDRS duration and F-M provoking factor) ratings.

TABLE 1. *Summary, internal consistency, and intraclass correlation coefficients of overall dystonia ratings for each rating scale*

	UDRS[a]	F-M[b]	GDS
Mean ± SD	19.0 ± 16.7	16.5 ± 17.3	17.6 ± 18.6
Range	(2.2–76.4)	(1.2–86.2)	(1.6–85.2)
Cronbach's α	0.93	0.89	0.91
Intraclass correlation coefficient	0.71	0.78	0.72

[a]Ten subjects had only 4 (instead of 5) ratings.
[b]Two subjects had only 4 (instead of 5) ratings.
UDRS, Unified Dystonia Rating Scale; F-M, Fahn-Marsden Scale; GDS, Global Dystonia Rating Scale.

RESULTS

Patients

A total of 103 patients were videotaped. One patient was excluded for failure to complete the videotape protocol, and 2 patients were excluded for having other neurological conditions besides primary dystonia. F-M data on 2 subjects from one rater and UDRS data on 8 subjects from another rater were missing; these data were excluded in analyses. Other isolated missing items were imputed in consultation with the PI.

There were 58 women and 42 men with primary dystonia included in the study. The patients had a mean age of 51 years (SD = 14.8). All forms of dystonia were represented (39 focal; 37 segmental and 24 generalized), and dystonic involvement of all body regions was represented. The mean ratings and range for each rating scale are shown in Table 1.

Internal Consistency

Each of the three scales was found to have a high level of internal consistency, with Cronbach's α ranging from 0.89 to 0.93 (Table 1). Cronbach's α is a function of the number of items on a rating scale and inter-rater correlation; it is an index of how stable and consistent the items on the scale are in measuring a single characteristic such as dystonia.

Inter-Rater Agreement

Each scale showed a high level of inter-rater reliability for the total scores, with the intraclass correlation coefficients ranging from 0.71 to 0.78 (Table 1). The results of the Kendall's coefficient of concordance for each body area for each scale are shown in Table 2. In general, the ratings for motor severity in the UDRS and the F-M showed higher levels of agreement than did the duration factor for the UDRS or the provoking factor from the F-M. The agreement is lowest for the larynx and speech for the UDRS (Kendall's = 0.56) and for the GDS (Kendall's = 0.59). Upper face and eyes showed the lowest agreement on the UDRS and the F-M.

TABLE 2. *Agreement of raters for motor severity ratings of different body regions: Kendall's coefficient of concordance*

	UDRS		F-M		
Body region	Severity	Duration	Severity	Provoking factor	GDS
Leg	0.76	0.73	0.75	0.72	0.75
Trunk	0.74	0.71	0.78	0.72	0.75
Arm	0.70	0.54	0.78	0.73	0.74
Neck	0.85	0.67	0.87	0.64	0.86
Larynx/speech	0.56	0.52	0.71	0.71	0.59
Lower face and jaw	0.68	0.64	0.71	0.65	0.73
Upper face and eyes	0.63	0.57	0.59	0.55	0.65

The generalized weighted κ statistic for each rating scale in each body area is shown in Table 3. In general, the motor severity ratings of both the UDRS and F-M showed greater agreement than the duration or provoking factors. As seen previously with the Kendall's, agreement among raters was lowest for upper face and eyes (motor severity of UDRS $\kappa = 0.52$; F-M $\kappa = 0.52$; GDS $\kappa = 0.58$).

Pairwise Comparisons and Pearson's Correlations

The total scores for the three scales are highly correlated with each other. The three pairwise scatterplots are shown in Figure 1. The scales had Pearson's correlations of 0.983 (UDRS and GDS), 0.977 (F-M and UDRS), and 0.980 (F-M and UDRS).

Investigator Questionnaires

The results of the investigator questionnaire are shown in Table 4. Seventy-four percent of the rating investigators found the GDS extremely or very easy to apply and 82% found it useful for measuring dystonia severity in an office setting. In contrast, only 5% of investigators found the UDRS easy to use, and 38% the F-M.

DISCUSSION

The measurement of dystonia severity lies with clinical examination and the development of reliable and valid rating scales. The testing of new therapeutic approaches to dystonia, including surgical interventions, will require collaboration among multiple investigators and study sites. To implement multicenter studies, a reliable and valid instrument that can assess the spectrum of dystonia severity is necessary.[9] The F-M dystonia rating scale has been used as the standard outcome measure.[10–12] Only one small study, however, has demonstrated reliability and validity of the F-M.[3] Whether this rating scale is useful for large clinical trials had not been assessed until the current study.

The UDRS was developed to address the perceived limitations of the F-M. The UDRS divides the body regions into smaller more defined areas, adding a new modifying rating scale that was successfully used for focal dystonia (TWSTRS),[4–6] and eliminates the weighting factor that lessened the contribution of dystonia severity in certain body regions in the F-M scale. The GDS was designed as a simple direct assessment of overall severity of dystonia in each body area. Despite differences in scale construction, the measures of internal consistency and inter-rater reliability were within acceptable range for all three scales. The Cronbach's α may be somewhat inflated because of the number of items within each scale, but the results suggest a stable construction of

TABLE 3. *Agreement of raters for motor severity ratings of different body regions: Generalized weighted κ*

	UDRS		F-M		
Body region	Severity	Duration	Severity	Provoking factor	GDS
Leg	0.87	0.80	0.91	0.85	0.80
Trunk	0.90	0.75	0.88	0.81	0.86
Arm	0.82	0.44	0.90	0.89	0.83
Neck	0.81	0.74	0.84	0.51	0.82
Larynx/speech	0.66	0.44	0.82	0.77	0.82
Lower face and jaw	0.63	0.49	0.62	0.61	0.73
Upper face and eyes	0.52	0.43	0.52	0.37	0.58

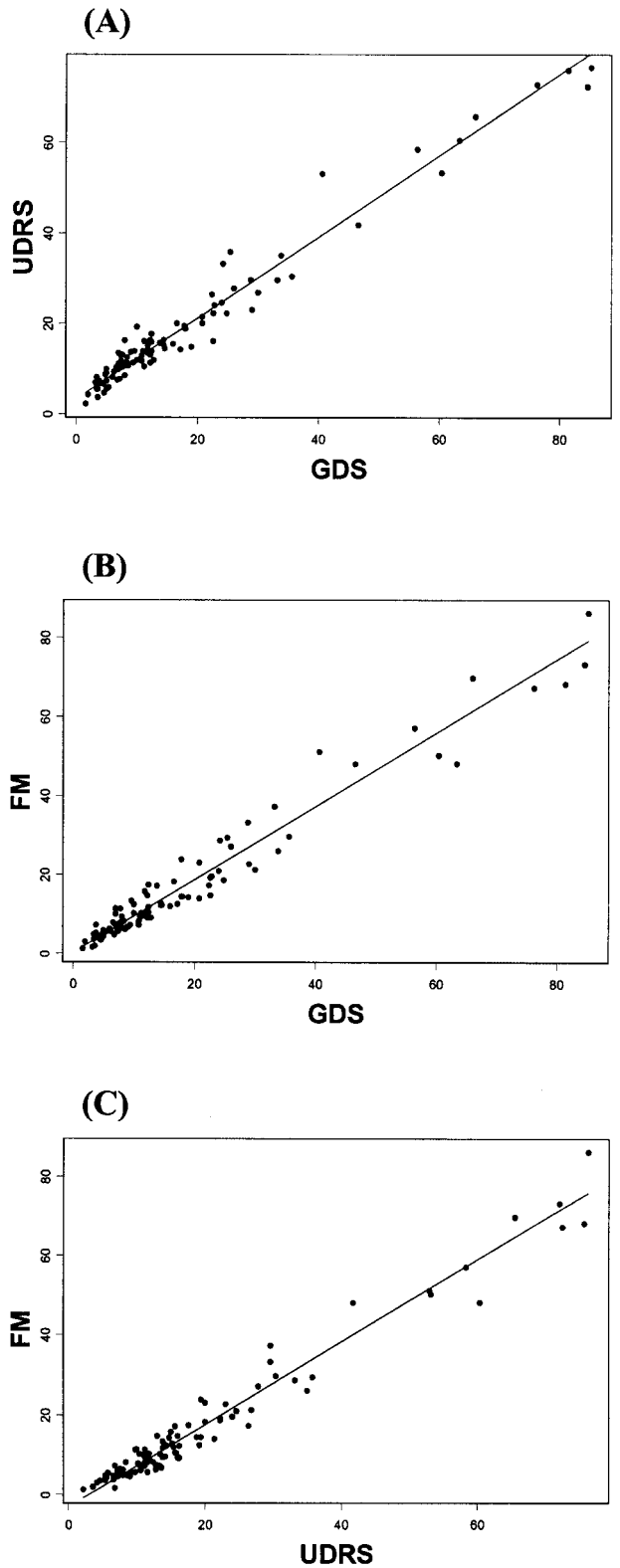

FIG. 1. Scatterplots assessing correlations of total scores between UDRS, F-M, and GDS. Pearson's *r* correlation coefficient.

TABLE 4. *Investigators' assessments of the ease of application and usefulness of each dystonia rating scale in clinical trials and office setting*

	UDRS	F-M	GDS
Extremely or very easy to apply	1/20 = 5%	8/21 = 38%	14/19 = 74%
Useful in multi-center clinical trials	19/20 = 90%	16/21 = 76%	8/22 = 36%
Useful in an office setting	9/21 = 43%	16/21 = 76%	18/22 = 82%

Percentage of investigators declaring specific characteristic of each scale, among those who replied.

these scales to assess dystonia. These results suggest that the items for each scale are consistently measuring the domain of interest.

The intra-class correlation coefficients demonstrate good to excellent inter-rater agreement for total scores on all three scales. This indicates that despite the individual differences that exist for items within a scale, raters assign the composite score in a similar manner.

The inter-rater agreement for individual items (body regions) on all three scales (F-M, UDRS, GDS) and for modifying ratings on two of the scales (F-M, UDRS) was fair to excellent using both the Kendall's coefficient of concordance and the generalized weighted κ statistic. The Kendall's coefficient of concordance provides an estimate of the consistency among raters for the rank order of ratings and the κ assesses the exact agreement in ratings.

The body regions showing the lowest level of inter-rater agreement were the lower face and jaw, and upper face and eyes. All three scales showed a similar pattern, suggesting difficulty in the assessment of dystonia in these particular anatomical areas. It may be that the distinction between frequent normal facial movements, such as eye blinks and facial expression, may be difficult to distinguish from mild intermittent dystonia. Alternatively, it may be that the videotape examination outlined in the protocol for these areas does not provide sufficient information for rating.

Likewise, the modifying ratings for the UDRS (Duration) and the F-M (Provoking Factor) showed consistently lower levels of agreement than motor severity ratings. The modifying factors are complex, combining presence of dystonia in particular situations, and assessment of maximal or submaximal intensity during the examination. The contribution of these modifying ratings to the reliability of the rating scales is modest, and the complexities of these additional ratings likely reduce the clinical usefulness of both the UDRS and the F-M. Although reliability and agreement seem equivalent among the three scales, the ease of application of the GDS reported by the majority of raters in this study suggests that this method for rating of dystonia se-

verity may be the most practical to implement in multiple research sites. As with rating scale development in other movement disorders, the next steps include an assessment of these scales for factor structure, revision of the scales with revisions to clarify rating items and possible deletion of the modifying rating of the UDRS and F-M. Tests for validity and responsivity to change are also necessary to understand the clinical utility of the scales. The parallel development of a teaching tape that demonstrates the ratings for each body area, especially face and eyes, will increase the inter-rater agreement for dystonia in these areas.

Acknowledgment: This work was supported by a grant from the Dystonia Medical Research Foundation.

APPENDIX 1

The following Dystonia Study Group sites and investigators participated in this study:

M. Stacy, Barrow Neurological Institute, Phoenix, AZ; D. Tarsy, Beth Israel Deaconess Medical Center, Boston, MA; J. Friedman, Boston Medical Center, Boston, MA; L. Seeberger, Colorado Neurological Institute, Englewood, CO; B. Ford, Columbia Presbyterian Medical Center, New York, NY; M. Evatt, Emory University, Atlanta, GA; O. Suchowersky, Foothills Hospital, Calgary, Canada; D. Riley, Hospital of Cleveland, Cleveland, OH; M. Jog, London Health Sciences Center, London, Ontario; M.F. Gordon, Long Island Jewish Hospital, New Hyde Park, NY; C. Adler, Mayo Clinic, Scottsdale, AZ; M. Brandabur, Neuropsych Institute, Chicago, IL; M. Hallett and B. Karp, NINDS, Bethesda, MD; S. Factor, Parkinson's Disease and Movement Disorders Center of Albany Medical Center, Albany, NY; D. Truong, The Parkinson's Disease and Movement Disorders Institute, Fountain Valley, CA; R. Chen, Toronto Western Hospital, Toronto, Canada; J. Tsui, University Hospital, Vancouver, Canada; U. Kang, University of Chicago, Chicago, IL; A. Brashear, University of Indiana, Indianapolis, IN; M. Swenson, University of Louisville, Louisville, KY; P. Tuite, University of Minnesota, Minneapolis, MN; M. Lew and G. Petzinger, University of Southern California, Los Angeles, CA; D. Trugman, University of Virginia Health Sciences Center, Charlottesville, VA.

APPENDIX 2. *Dystonia Study Group Videotape examination protocol*

Area assessed	Perspective	Activity
Part 1: Eyes and upper face	Close view of head and shoulders; sitting unsupported in chair without back	At rest (10 sec) Eyes open (10 seconds close view, 10 seconds far view) Eyes closed (10 seconds close view, 10 seconds far view) Forced eye blinks: 10 repetitions (10 sec)
Part 2: Lower face, jaw, tongue, larynx	Patient seated	Close view of face at rest (10 sec) Reading: standardized passage aloud (Rainbow passage). First 3 lines Repeated consonants: Tee, Mee, La, Ca: 5 of each (15 sec) Holding the note "eeee" for 5 seconds Count to 10 (5 sec) Tongue protrusion: (5 sec) Opening and closing mouth for 5 reps (10 sec)
	Swallow interview	Question to patient: Do you have problems with swallowing? If yes, is it occasional or frequent? Do you choke occasional or frequently? Can you swallow firm foods? Liquids?
Part 3: Neck	Seated in chair, close view head and shoulders	Frontal view at rest (instruct to allow head to move) (10 sec) Seated with eyes closed (instruct to allow head to move) (10 sec) Quiet conversation for 2 sentences (10 sec) Turn head all the way to right then left Tilt ear to shoulder on each side Look up and look down Lateral view (5 sec) Walking back and forth twice (total 20 sec)
Part 4: Shoulders and upper arms, distal arm and hands	Far view of upper half of body	Arms extended supinated: 5 sec Arms extended pronated: 5 sec Arms flexed at elbow in front of chest: 5 sec Finger to nose: 5 repetitions (5 sec) Finger tapping, right than left: 5 reps (5 sec) Flex and extend wrists with arms outstretched for 5 reps (5 sec) Cup to lips, right than left arm (5 sec) Writing: "Today is a nice day" for 3 repetitions (maximum time 15 sec) Drawing spiral without hand resting on paper; right than left hand (maximum time 10 sec) Hold up spiral

APPENDIX 2. (*Continued*)

Area assessed	Perspective	Activity
Part 5: Upper leg, distal leg, foot and trunk	Far view entire body, sitting	Sitting quietly (10 sec)
		Heel to toe taps: 5 reps on each side (10 sec)
	Far view entire body: standing and walking	Standing frontal view for 10 sec
		Standing: lateral view for 5 sec
		Standing: back view for 5 sec
		Walking: away and toward examiner 20 feet: 2 reps (maximum 20 seconds)

APPENDIX 3: *Unified Dystonia Rating Scale (UDRS)*

Factor/area	Criteria
Duration	
0	None
0.5	Occasional (<25% of the time); predominantly submaximal
1.0	Occasional (<25% of the time); predominantly maximal
1.5	Intermittent (25–50% of the time); predominantly submaximal
2.0	Intermittent (25–50% of the time); predominantly maximal
2.5	Frequent (50–75% of the time); predominantly submaximal
3.0	Frequent (50–75% of the time); predominantly maximal
3.5	Constant (>75% of the time); predominantly submaximal
4.0	Constant (>75% of the time); predominantly maximal
Motor severity	
Eyes and upper face	
0	None
1	Mild: increased blinking or slight forehead wrinkling (≤25% maximal intensity)
2	Moderate: eye closure without squeezing or pronounced forehead wrinkling (>25% but ≤50% maximal intensity)
3	Severe: eye closure with squeezing, able to open eyes within 10 seconds or marked forehead wrinkling (>50% but ≤75% maximal intensity)
4	Extreme: eye closure with squeezing, unable to open eyes within 10 seconds or intense forehead wrinkling (>75% maximal intensity)
Lower face	
0	None
1	Mild: grimacing of lower face with minimal distortion of mouth (≤25% maximal)
2	Moderate: grimacing of lower face with moderate distortion of mouth (>25% but ≤50% maximal)
3	Severe: marked grimacing with severe distortion of mouth (>50% but ≤75% maximal)
4	Extreme: intense grimacing with extreme distortion of mouth (>75% maximal)
Jaw and tongue	
0	None
1	Mild: jaw opening or tongue protrusion ≤25% of possible range or forced jaw clenching without bruxism
2	Moderate: jaw opening or tongue protrusion >25% but ≤50% of possible range or forced jaw clenching with mild bruxism secondary to dystonia
3	Severe: jaw opening and/or tongue protrusion >50% but ≤75% of possible range or forced jaw clenching with pronounced bruxism secondary to dystonia
4	Extreme: jaw opening or tongue protrusion >75% of possible range or forced jaw clenching with inability to open mouth
Larynx	
0	None
1	Mild: barely detectable hoarseness or choked voice or occasional voice breaks
2	Moderate: obvious hoarseness or choked voice or frequent voice breaks
3	Severe: marked hoarseness or choked voice or continuous voice breaks
4	Extreme: unable to vocalize
Neck	
0	None
1	Mild: movement of head from neutral position ≤25% of possible normal range
2	Moderate: movement of head from neutral position >25% but ≤50% of possible normal range
3	Severe: movement of head from neutral position >50% but ≤75% of possible normal range
4	Extreme: movement of head from neutral position >75% of possible normal range

APPENDIX 3: (*Continued*)

Factor/area	Criteria
Shoulder and proximal arm (right and left)	
0	None
1	Mild: movement of shoulder or upper arm ≤25% of possible normal range
2	Moderate: movement of shoulder or upper arm 25% but ≤50% of possible normal range
3	Severe: movement of shoulder or upper arm 50% but ≤75% of possible normal range
4	Extreme: movement of shoulder or upper arm 75% of possible normal range
Distal arm and hand including elbow (right and left)	
0	None
1	Mild: movement of distal arm or hand ≤25% of possible normal range
2	Moderate: movement of distal arm or hand 25% but ≤50% of possible normal range
3	Severe: movement of distal arm or hand 50% but ≤75% of possible normal range
4	Extreme: movement of distal arm or hand 75% of possible normal range
Pelvis and proximal leg (right and left)	
0	None
1	Mild: tilting of pelvis or movement of proximal leg or hip ≤25% of possible normal range
2	Moderate: tilting of pelvis or movement of proximal leg or hip 25% but ≤50% of possible normal range
3	Severe: tilting of pelvis or movement of proximal leg or hip 50% but ≤75% of possible normal range
4	Extreme: tilting of pelvis or movement of proximal leg or hip 75% of possible normal range
Distal leg and foot including knee (right and left)	
0	None
1	Mild: movements of distal leg or foot ≤25% of possible normal range
2	Moderate: movements of distal leg or foot 25% but ≤50% of possible normal range
3	Severe: movements of distal leg or foot 50% but ≤75% of possible normal range
4	Extreme: movements of distal leg or foot 75% of possible normal range
Trunk	
0	None
1	Mild: bending of trunk ≤25% of possible normal range
2	Moderate: bending of trunk 25% but ≤50% of possible normal range
3	Severe: bending of trunk >50% but ≤75% of possible normal range
4	Extreme: bending of trunk >75% of possible normal range

APPENDIX 4A. *Fahn Marsden rating scale*

Region	Provoking factor	Severity factor	Weight	Product
Eyes	0–4	×0–4	0.5	0–8
Mouth	0–4	×0–4	0.5	0–8
Speech and swallow	0–4	×0–4	1.0	0–16
Neck	0–4	×0–4	0.5	0–8
Arm (R)	0–4	×0–4	1.0	0–16
Arm (L)	0–4	×0–4	1.0	0–16
Trunk	0–4	×0–4	1.0	0–16
Leg (R)	0–4	×0–4	1.0	0–16
Leg (L)	0–4	×0–4	1.0	0–16
Sum				Max 120

APPENDIX 4B. *Fahn Marsden rating factors*

Factor/area/rating	Criteria
I. Provoking factor	
General	
0	No dystonia at rest or with action
1	Dystonia only with particular action
2	Dystonia with many actions
3	Dystonia on action of distant part of body or intermittently at rest
4	Dystonia present at rest
Speech and swallowing	
1	Occasional, either or both
2	Frequent either
3	Frequent one and occasional other
4	Frequent both
II. Severity factor	
Eyes	
0	No dystonia
1	Slight: Occasional blinking
2	Mild. Frequent blinking without prolonged spasms of eye closure
3	Moderate. Prolonged spasms of eyelid closure, but eyes open most of the time
4	Severe. Prolonged spasms of eyelid closure, with eyes closed at least 30% of the time
Mouth	
0	No dystonia present
1	Slight. Occasional grimacing or other mouth movements (e.g., jaw opened or clenched; tongue movement
2	Mild. Movement present less than 50% of the time
3	Moderate dystonic movements or contractions present most of the time
4	Severe dystonic movements or contractions present most of the time
Speech and swallowing	
0	Normal
1	Slightly involved; speech easily understood or occasional choking
2	Some difficulty in understanding speech or frequent choking
3	Marked difficulty in understanding speech or inability to swallow firm foods
4	Complete or almost complete anarthria, or marked difficulty swallowing soft foods and liquids
Neck	
0	No dystonia present
1	Slight. Occasional pulling
2	Obvious torticollis, but mild
3	Moderate pulling
4	Extreme pulling
Arm	
0	No dystonia present
1	Slight dystonia. Clinically insignificant
2	Mild: Obvious dystonia, but not disabling
3	Moderate. Able to grasp, with some manual function
4	Severe. No useful grasp
Trunk	
0	No dystonia present
1	Slight bending; clinically insignificant
2	Definite bending, but not interfering with standing or walking
3	Moderate bending; interfering with standing or walking
4	Extreme bending of trunk preventing standing or walking
Leg	
0	No dystonia present
1	Slight dystonia, but not causing impairment; clinically insignificant
2	Mild dystonia. Walks briskly and unaided
3	Moderate dystonia. Severely impairs walking or requires assistance
4	Severe. Unable to stand or walk on involved leg

APPENDIX 5

The Global Dystonia Severity Rating Scale

The global score is an overall score for the body area. The investigator rates the patient in relationship to all patients. If the dystonia changes during the examination, the rating for the maximal dystonia is recorded.

Each body area is rated from 0 to 10:

0 No dystonia present in that body area
1 Minimal dystonia
5 Moderate dystonia
10 Most severe dystonia

Ten body areas are tested: 1) Eyes and upper face, 2) lower face, 3) jaw and tongue, 4) larynx, 5) neck, 6) shoulder and proximal arm, 7) distal arm and hand including elbow, 8) pelvis and upper leg, 9) distal leg and foot, and 10) trunk.

REFERENCES

1. Fahn S, Marsden CD, Calne DB. Classification and investigation of dystonia. In: Marsden CD, Fahn S, editors. Movement disorders 2. London: Butterworth and Co.; 1987. p 332–358.
2. Fahn S. Assessment of the primary dystonias. In: Munsat TL, editor. Quantification of neurological deficit. London: Butterworth; 1989. p 241–270.
3. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. Neurology 1985;35:73–77.
4. Consky ES, Basinki A, Belle L, Ranawaya R, Lang AE. The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS): assessment of the validity and inter-rater reliability. Neurology 1990;40(Suppl):445.
5. Consky ES, Lang AE. Clinical assessments of patients with cervical dystonia. In: Jankovic J, Hallett M, editors. Therapy with botulinum toxin. New York: Marcel Dekker, Inc.; 1994. p 211–237.
6. Comella CL, Stebbins GT, Goetz CG, Chmura T, Bressman SB, Lang AE. Teaching tape for the motor section of the Toronto Western Spasmodic Torticollis Scale. Mov Disord 1997;12:570–575.
7. Armitage P, Berry G. Statistical methods in medical research, Third ed. Oxford: Blackwell Science Ltd.; 1994.
8. Fleiss JL. Statistical methods for rates and proportions, Second ed. New York: Wiley; 1981.
9. Hobart JC, Lamping DL, Thompson AJ. Evaluating neurological outcome measures: the bare essentials. J Neurol Neurosurg Psychiatry 1996;60:127–130.
10. Burke RE, Fahn S, Marsden CD. Torsion dystonia: a double-blind, prospective trial of high-dosage trihexyphenidyl. Neurology 1986; 36:160–164.
11. Volkman J, Benecke R. Deep stimulation for dystonia: patient selection and evaluation. Mov Disord 2002;17(Suppl):S112–S115.
12. Vercueil L, Krack P, Pollak P. Results of deep brain stimulation for dystonia: a critical reappraisal. Mov Disord 2002;17(Suppl):S89–S93.
13. Fahn S, Elton RL, and members of the UPDRS development committee. Unified Parkinson's disease rating scale. In: Fahn S, Marsden CD, Calne DB, Goldstein M, editors. Recent development in Parkinson's disease. Vol 2. Florham Park, NJ: Macmillan Health Care Information; 1987. p 153–164.
14. Goetz CG, Stebbins GT, Shale HM, Lang AE, Chernik DA, Chmura TA, Ahlskog JE, Dorflinger EE. Utility of an objective dyskinesia rating scale for Parkinson's disease: inter- and intra-rater reliability assessment. Mov Disord 1994;9:390–394.
15. Huntington Study Group. Unified Huntington's disease rating scale: reliability and consistency. Mov Disord 1996;11:136–142.
16. Stebbins GT, Goetz CG, Flournoy T. Unified Parkinson's disease rating scale: reliability and factorial validity of the motor exam section. Ann Neurol 1991;30:298.
17. Goetz CG, Stebbins GT, Chmura TA, Fahn S, Klawans HL, Marsden CD. Teaching tape for the motor section of the Unified Parkinson's disease rating scale. Mov Disord 1995;10:263–266.