For quantitative assessment of the primary torsion dystonias, a rating scale is proposed that has two sections—a Movement Scale, based on examination, and a Disability Scale, based on the patient’s statements about seven activities of daily living. We assessed the validity of the Movement Scale by comparing scores with a ranking of patients according to dystonia severity and with ratings of the patients on the Disability Scale. In addition, we assessed the inter- and intra-rater reliability of the scale by comparing independent scorings of patients by four examiners and by comparing scorings by the same examiners performed at different times. We found that the Movement Scale was a valid and reliable indicator of the severity of primary torsion dystonia.

Validity and reliability of a rating scale for the primary torsion dystonias

Robert E. Burke, Stanley Fahn, C. David Marsden, Susan B. Bressman, Carol Moskowitz, and Joseph Friedman

Progress in treating any neurologic disease, as well as in characterizing the clinical course, depends on an accurate and practical means of assessing severity. Reliable assessment scales have been developed for Parkinson's disease and tardive dyskinesia, but not for many other movement disorders. Torsion dystonia has not been assessed with quantitative scales, partly because it is a complex movement disorder that affects different parts of the body in different ways and often changes in the same patient at different times.

Fahn and Marsden proposed a clinical assessment scale for primary torsion dystonia based on an examination of the patient in a standard setting and on the patient’s subjective report of disability in activities of daily living. This scale was developed for a therapeutic trial of trihexyphenidyl in the treatment of dystonia. We now present a complete description of the scale and a study of its validity and reliability. If the scale is valid, increasing scores should correlate with both clinical impression of the severity of dystonia and with increasing disability in the activities of daily living. If the scale is reliable, the same examiner should obtain the same score at different times if the disorder has not changed clinically between the two examinations (intra-rater reliability). In addition, different examiners of the same patient should obtain similar scores (inter-rater reliability). We examined the validity and reliability of this scale using videotapes of patients with primary dystonia, filmed according to a standard format.

Methods. We define dystonia as an involuntary movement disorder characterized by twisting or sustained movements. The Fahn-Marsden scale has two sections: a Movement Scale, based on examination of the patient (table 1) and a Disability Scale, based on the patient’s view of disability in activities of daily living (table 2).

The Movement Scale score is the sum of individual scores for each of nine body “regions” (speech and swallowing are considered together as a region). The individual score for each region is the product of two factors—the Provoking Factor and the Severity Factor, each rated from 0 (lowest) to 4 (highest).

The Provoking Factor quantifies the dystonia in a given region by rating the circumstances in which dystonia appears. The most severe state is persistent dystonia, noted even when the patient is sitting at rest (Provoking Factor = 4). Somewhat less severe is dystonia that appears intermittently while the patient sits or dystonia in the region being rated when some other area of the body is performing an action ("overflow") (Factor = 3). An example of overflow is the appearance of dystonic movement in the feet when the patient opens and closes the hands voluntarily. Still less severe is dystonia that appears only when the region rated is being used and with only one particular action (Factor = 1), such as dystonia in the hand only during the act of writing or dystonia in the feet only on walking. Because speech and swallowing
Table 1. Dystonia movement scale

<table>
<thead>
<tr>
<th>Region</th>
<th>Provoking factor</th>
<th>Severity factor</th>
<th>Weight</th>
<th>Product</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eyes</td>
<td>0-4</td>
<td>0-4</td>
<td>0.5</td>
<td>0-8</td>
</tr>
<tr>
<td>Mouth</td>
<td>0-4</td>
<td>0-4</td>
<td>0.5</td>
<td>0-8</td>
</tr>
<tr>
<td>Speech/ swallowing</td>
<td>0-4</td>
<td>0-4</td>
<td>1.0</td>
<td>0-16</td>
</tr>
<tr>
<td>Neck</td>
<td>0-4</td>
<td>0-4</td>
<td>0.5</td>
<td>0-8</td>
</tr>
<tr>
<td>R arm</td>
<td>0-4</td>
<td>0-4</td>
<td>1.0</td>
<td>0-16</td>
</tr>
<tr>
<td>L arm</td>
<td>0-4</td>
<td>0-4</td>
<td>1.0</td>
<td>0-16</td>
</tr>
<tr>
<td>R leg</td>
<td>0-4</td>
<td>0-4</td>
<td>1.0</td>
<td>0-16</td>
</tr>
<tr>
<td>L leg</td>
<td>0-4</td>
<td>0-4</td>
<td>1.0</td>
<td>0-16</td>
</tr>
</tbody>
</table>

Sum: (maximum = 120)

I. Provoking factor

A. General

0 - No dystonia at rest or with action
1 - Dystonia on particular action
2 - Dystonia on many actions
3 - Dystonia on action of distant part of body or intermittently at rest
4 - Dystonia present at rest

B. Speech and swallowing

1 - Occasional, either or both
2 - Frequent either
3 - Frequent one and occasional other
4 - Frequent both

II. Severity factors

Eyes

0 - No dystonia present
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 open 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
Table 2. Disability scale

<table>
<thead>
<tr>
<th>Function</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech</td>
<td>0-4</td>
</tr>
<tr>
<td>Writing</td>
<td>0-4</td>
</tr>
<tr>
<td>Feeding</td>
<td>0-4</td>
</tr>
<tr>
<td>Eating</td>
<td>0-4</td>
</tr>
<tr>
<td>Hygiene</td>
<td>0-4</td>
</tr>
<tr>
<td>Dressing</td>
<td>0-4</td>
</tr>
<tr>
<td>Walking</td>
<td>0-6</td>
</tr>
</tbody>
</table>

Sum: (Maximum = 30)

Disability scale

A. Speech
0 - Normal 1 - Slightly involved; easily understood 2 - Some difficulty in understanding 3 - Marked difficulty in understanding 4 - Complete or almost complete anarthria

B. Handwriting (tremor or dystonia)
0 - Normal 1 - Slight difficulty; legible 2 - Almost illegible 3 - Illegible 4 - Unable to grasp to maintain hold on pen

C. Feeding
0 - Normal 1 - Uses "tricks"; independent 2 - Can feed, but not cut 3 - Finger food only 4 - Completely dependent

D. Eating/swallowing
0 - Normal 1 - Occasional choking 2 - Chokes frequently; difficulty swallowing 3 - Unable to swallow firm foods 4 - Marked difficulty swallowing soft foods and liquids

E. Hygiene
0 - Normal 1 - Clumsy; independent 2 - Needs help with some activities 3 - Needs help with most activities 4 - Needs help with all activities

F. Dressing
0 - Normal 1 - Clumsy, independent 2 - Needs help with some activities 3 - Needs help with most activities 4 - Helpless

G. Walking
0 - Normal 1 - Slightly abnormal; hardly noticeable 2 - Moderately abnormal; obvious to naive observer 3 - Considerably abnormal 4 - Needs assistance to walk 6 - Wheelchair-bound

are considered together as one region, the Provoking Factor score there differs from that of other regions, based on the patient’s report about how frequently either or both are involved (table 1). A Provoking Factor of 1 is selected if there is only occasional (<1 episode per month) difficulty with speech, swallowing, or both. If either is frequently affected (>1 episode per month—eg, of choking), a Provoking Factor of 2 is chosen. If one is frequently affected and the other only occasionally, the Factor is 3; if both are frequently affected, the Factor is 4.

The Severity Factor quantifies the severity of dystonia in a region regardless of the circumstances in which dystonia appears. The 0 to 4 rating of severity for each region is defined explicitly for that region (table 1). For example, involvement of the trunk is maximum when extreme bending prevents standing or walking (Severity Factor = 4). Somewhat less severe is bending that interferes with standing or walking, but does not prevent it entirely (Factor = 3). Still less severe is definite bending that never causes loss of balance sufficient to interfere with standing or walking (Factor = 2). The least severe is detectable bending that is mild, sometimes not even noted by the patient ("clinically insignificant").

After each region is rated for Provoking Factor and Severity Factor, the two are multiplied to give a product for that region. For the eyes, mouth, and neck, the product is further multiplied by 0.5 to "down weight" the scores for those regions, because their involvement seems to add less to the overall disability. A maximum Movement Scale score is 120; minimum is 0.

Some confusion could occur when dystonia affects shoulder or pelvic girdles, which are at junctions of at least two regions and could be considered to belong to any of them. The shoulder girdle could be a part of
the neck, trunk, or arm; the pelvic girdle, a part of the trunk or leg. We have considered dystonic movements of the trapezius muscle as part of the neck region. The trapezius is frequently involved in torticollis and is innervated by the accessory nerve, which also innervates the sternocleidomastoid muscle; we therefore classify trapezius involvement with nuchal dystonia. If shoulder girdle posturing displaces the arm (eg, internal or external rotations), we assign it to the arm region. If shoulder movements accompany kyphosis or scoliosis, we list the dystonia as part of the trunk region.

Tortipelvis is usually a feature of trunk dystonia, but if pelvic dystonia displaces the leg (eg, abduction, adduction, or rotation), it is considered to belong to the leg region.

For the Movement Scale examination, the patient sits with elbows and hands resting on the arms of the chair or on the thighs. In this position, any region with persistent dystonia is given a Provoking Factor score of 4. Any region with intermittent dystonia is given a score of 3. Although the neck and trunk are not truly at rest while the patient sits (because these muscles are working against gravity) they are nevertheless given a score of 4 if dystonia is apparent. The patient is now asked to perform standard acts, and the examiner determines whether dystonia is present in the region performing the act or in any other region. Appearance of dystonia in other regions is overflow. Particular actions of the individual regions is calculated to provide a convenient score, it is then assigned a Severity Factor by reference to the guidelines (table 1). The sum of the products of the individual regions is calculated to provide the Movement Scale score.

The Disability Scale score is the sum of individual ratings based on guidelines for seven activities of daily living (table 2). A maximum severity score is 30.

For validity and reliability studies, we used 10 conveniently available patients with primary dystonia who were part of a trihexyphenidyl trial, for which they had been assessed with the Dystonia Movement Scale and videotaped (table 3). Without reference to these previously recorded Scale scores, the patients were rated by three examiners according to the global severity of dystonia as seen on videotape. Each patient was rated by each examiner as having 1 to 5 (most severe) global dystonia involvement. The 10 patients were then ranked according to the sum of the global impression scores assigned by the three examiners. The patients on the videotapes were then scored independently by three trained examiners using the Dystonia Movement Scale. Another neurologist ("untrained examiner") had received no previous training in the use of the scale. On the day he used the scale to rate the videotaped patients, its use was explained to him.

To assess the validity of the Movement Scale, we evaluated correlations between patient scores and the global impression ranking, as well as between patient scores and Disability Scale scores. We used two tests to assess intra-rater reliability for two examiners. One examiner had previously scored the 10 patients "live"; those scores were compared with later scoring of videotapes of the same patients. A second trained examiner scored the 10 videotaped patients once and then again months later. Intra-rater reliability was assessed by examining correlations between scores of the tests of the 10 videotaped patients, performed independently by three examiners (R.E.B, S.B.B., C.M.) trained in the use of the

<table>
<thead>
<tr>
<th>Table 3. Videotape protocol</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1. Sitting at rest, arms on legs</strong></td>
</tr>
<tr>
<td>a. Whole body</td>
</tr>
<tr>
<td>b. Zoom in to different body regions</td>
</tr>
<tr>
<td>(head and neck, each hand, trunk, each foot)</td>
</tr>
<tr>
<td><strong>2. Speak—name, date, describe speech,</strong></td>
</tr>
<tr>
<td>swallowing, and current problems</td>
</tr>
<tr>
<td>a. Film whole body</td>
</tr>
<tr>
<td>b. Zoom in to different body parts</td>
</tr>
<tr>
<td><strong>3. Arms suspended in front of body—15 sec</strong></td>
</tr>
<tr>
<td>Finger-to-nose 5 times</td>
</tr>
<tr>
<td>Rapid succession movements: each hand and foot</td>
</tr>
<tr>
<td><strong>4. Arise and stand; turn 90° 4 times</strong></td>
</tr>
<tr>
<td>5. Walk</td>
</tr>
<tr>
<td>a. Whole body</td>
</tr>
<tr>
<td>b. Zoom in to different body regions</td>
</tr>
<tr>
<td><strong>6. Write with each hand</strong></td>
</tr>
<tr>
<td>Name, date, sentence, spiral</td>
</tr>
<tr>
<td>Videotape whole body and zoom in</td>
</tr>
<tr>
<td><strong>Total</strong></td>
</tr>
</tbody>
</table>
scale can be used to assess secondary dystonias, but dystonia is diagnosed when the neurologic examination, copper studies, analysis of CSF, or CT. The additionally, no abnormalities on slit-lamp examination reveals only dystonia; there is no history of birth injury, head injury, encephalitis, stroke, antipsychotic drug ingestion, or other causative factor associated with symptomatic dystonia; there are, additionally, no abnormalities on slit-lamp examination, copper studies, analysis of CSF, or CT. The scale can be used to assess secondary dystonias, but the scores are sometimes less meaningful. For example, a child with dystonia due to cerebral palsy may also have limb ataxia, weakness, and spasticity that cause inability to grasp or stand. Consequently, a noninterpretive use of the dystonia scale could give high scores even when there is little dystonia. On the other hand, deciding which disabilities are due to dystonia, and to what extent, is subjective and possibly unreliable.

An additional limitation of the scale is that it was designed to offer a broad range of scores to assess quantitatively different degrees of generalized dystonia. It is therefore insensitive to changes in focal dystonia; major changes are required before the change registers on the scale.

Nevertheless, we have found the scale most useful in providing valid, reliable documentation of the course of primary dystonia patients, particularly in response to therapy.

**Acknowledgment**

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**References**