



Scales to measure Dystonia

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The following disease groups are covered by ERN-RND:

- Ataxias and Hereditary Spastic Paraplegias
- Atypical Parkinsonism and genetic Parkinsons' Disease
- Dystonia, Paroxysmal Disorder and Neurodegeneration with Brain Ion Accumulation
- Frontotemporal Dementia
- Huntingtons' Disease and other Chorea
- Leukodystrophies

Specific information about the network, the expert centres and the diseases covered can be found at the networks web site www.ern-rnd.eu.

Recommendation for clinical use:

The European Reference Network for Rare Neurological Diseases strongly recommends the use the following scale as best clinical practice for the assessment and rating of Ataxia.



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METHODOLOGY

The development of the Diagnostic Flowcharts for Dystonia was done by the Disease group for Dystonia, Paroxysmal Disorder and NBIA of ERN-RND. Scales used in the clinical practice of the disease group members were mapped, and the decision on which scale should be proposed was taken by anonymous majority voting.

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Endorsement process:

- Mapping of used disease scales by disease group – 06 - 12/2018
- Survey to decide on scales by anonymous majority voting – 31.01. – 24.02.2019
- Consent on document by whole disease group – 26.03.2019



Recommended scales:

Domain	Scale
Generalised dystonia	Fahn-Marsden Dystonia Rating Scale ¹
Generalised dystonia in children	The Movement Disorder-Childhood Rating Scale ²
Blepharospasm	Blepharospasm severity rating scale ²
Cervical Dystonia	Toronto Western Spasmodic Torticollis Rating Scale for cervical dystonia ^{4,5}
Laryngeal Dystonia	Vocal Performance Questionnaire (VPQ) ⁶



Fahn-Marsden Dystonia Rating Scale¹

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



The Movement Disorder-Childhood Rating Scale²

Appendix 1. Movement Disorder Childhood Rating Scale for ages 4-18 years

Classification of MD	Prevalent MD*	Other MD
Hypokinetic-rigid syndrome	<input type="checkbox"/>	<input type="checkbox"/>
Chorea/ballism	<input type="checkbox"/>	<input type="checkbox"/>
Dystonia/athetosis	<input type="checkbox"/>	<input type="checkbox"/>
Myoclonus	<input type="checkbox"/>	<input type="checkbox"/>
Tic	<input type="checkbox"/>	<input type="checkbox"/>
Tremor	<input type="checkbox"/>	<input type="checkbox"/>

Part I. General Assessment

A. Motor function

1. Head control

- 0 normal
- 1 mildly affected by the MD and/or control longer than 1 min
- 2 moderately affected by the MD and/or control between 30 seconds and 1 min
- 3 severely affected by the MD and/or control shorter than 30 seconds
- 4 absent

2. Sitting position

- 0 normal
- 1 mildly affected by the MD and/or control longer than 1 min
- 2 moderately affected by the MD and/or control between 30 seconds and 1 min
- 3 severely affected by the MD and/or control shorter than 30 seconds
- 4 absent

3. Standing position

- 0 normal
- 1 maintained for >1 min, even with abnormal posture
- 2 maintained for >30 seconds and <1 min, even with abnormal posture
- 3 maintained for <30 seconds, or for long periods with support
- 4 absent or for brief periods with support

4. Walking

- 0 normal
- 1 walks alone for >5 min, even with abnormal posture
- 2 walks alone for <5 min, even with abnormal posture or needs support
- 3 only a few steps with support, and with very abnormal pattern
- 4 absent

5. Reaching

- 0 normal
- 1 possible for age-adequate tasks, even if qualitatively abnormal
- 2 moderately abnormal, only possible to approach the object
- 3 severely abnormal, only inefficient efforts
- 4 absent

6. Grasping

- 0 normal
- 1 mildly abnormal, but pincer-grasp possible
- 2 moderately abnormal, only possible for cubes very close to the hand
- 3 severely abnormal, only inefficient efforts
- 4 absent

7. Handwriting

- 0 normal
- 1 mild difficulties, but readable
- 2 moderate difficulty, not completely readable
- 3 severe difficulties, not readable
- 4 absent, a functional grasp of the pen is not possible



Blepharospasm severity rating scale³

Intensity rating

ITEM A1) Type of eyelid spasm

- Brief (< 3 sec duration) eyelid spasms with complete rim closure = score 1
- Prolonged (\geq 3 sec duration) eyelid spasms with partial rim closure = score 2
- Prolonged (\geq 3 sec duration) eyelid spasms with complete rim closure = score 3

ITEM A2) Apraxia of eyelid opening

- Yes = score 2
- No = score 0

ITEM A3) OO spasms occur during writing:

- Yes = score 1
- No = score 0

ITEM A4) Average duration of prolonged eyelid spasm with complete rim closure recorded while patient at rest, eyes open, for two minutes. Calculate the correspondent tertile as follows:

- I tertile = 3 to 4 sec = score 1
- II tertile = 4.1 to 5 sec = score 2
- III tertile = > 5 sec = score 3

Frequency rating

ITEM B1) Count “number of blinks + brief eyelid spasm / min” (patient at rest, eyes open, for two minutes) and calculate the corresponding tertile as follows:

- I tertile = 1 – 18 blinks + brief spasm / min = score 1
- II tertile = 19 – 32 blinks + brief spasm /min = score 2
- III tertile = > 32 blinks + brief spasm / min = score 3

ITEM B2) Count number of prolonged eyelid spasm with complete rim closure / min” (patient at rest, eyes open, for two minutes) and calculate the corresponding quartile as follows:

- I tertile = 1 – 3/ min = score 1
- II tertile = 3.1 – 7/min = score 2
- III tertile = > 7/ min = score 3

Total score = Intensity + Frequency = (A1 + A2 + A3 + A4) + (B1 + B2)



Toronto Western Spasmodic Torticollis Rating Scale for cervical dystonia^{4,5}

This scale is used to assess the severity of cervical dystonia and the success of its treatment. A total score of 0 to 35 can be achieved; this is made up of various subscores (A-F).

A. Maximal excursion		
Assessment of the maximum range of movement. The patient is asked not to suppress the involuntary movement. The examiner can use provocation manoeuvres which make the symptoms worse. If the severity lies between two values, the value for the more severe disorder is selected.		
1. Rotation (left or right)	0	None
	1	Slight (1-22°)
	2	Mild (23-45°)
	3	Moderate (46-67°)
	4	Severe (68-90°)
2. Laterocollis (excluding shoulder elevation)	0	None
	1	Mild (1-15°)
	2	Moderate (16-35°)
	3	Severe (>35°)
3. Anterocollis/retrocollis	a) Anterocollis	
	0	None
	1	Mild downward deviation of chin
	2	Moderate downward deviation of chin
	3	Severe (chin reaches chest)
	b) Retrocollis	
	0	None
	1	Mild backward deviation
	2	Moderate (half the possible range of movement)
	3	Severe
4. Lateral shift	0	Absent
	1	Present
5. Sagittal shift	0	Absent
	1	Present



B. Duration (weighted x 2)	
0	None
1	Occasional deviation (<25% of the time, most often submaximal)
2	Occasional deviation (<25% of the time, most often maximal) or intermittent deviation (25-50% of the time, most often submaximal)
3	Intermittent deviation (25-50% of the time, often maximal) or frequent deviation (50-75% of the time, most often submaximal)
4	Frequent deviation (50-75% of the time, most often maximal) or constant deviation (>75% of the time, most often submaximal)
5	Constant deviation (>75% of the time, often maximal)
C. Effect of sensory tricks	
0	Complete relief
1	Partial or only limited relief
2	Little or no benefit
D. Shoulder elevation or anterior displacement	
0	Absent
1	Mild (<1/3 of possible range), intermittent or moderate and intermittent
2	Moderate (1/3 to 2/3 of possible range and constant, >75% of the time) or severe (>2/3 of possible range) and intermittent
3	Severe and constant
E. Range of motion (without sensory tricks)	
If there are limitations on more than one level, the highest value is recorded in each case.	
0	Able to move to extreme opposite position
1	Able to move head well past midline but not to extreme opposite position
2	Able to move head barely past midline
3	Able to move head toward but not past midline
4	Barely able to move head beyond abnormal posture
F. Time	
Maximum time for which patient is able to maintain head in neutral position without sensory tricks	
0	>60 sec
1	46-60 sec



2	31-45 sec
3	16-30 sec
4	<15 sec



Vocal Performance Questionnaire⁶

Appendix 1. Vocal Performance Questionnaire

By Paul Carding, Freeman Hospital, Newcastle upon Tyne, UK

Name Date

Tick or circle an answer for each question.

- 1 How do you think your voice sounds now (compared with before your voice problems started)?
 - (a) No different from usual voice
 - (b) Only slightly different from usual voice
 - (c) Quite different from usual voice
 - (d) Very different from usual voice
 - (e) Totally different from usual voice
- 2 Does your voice give you any physical discomfort when you talk?
 - (a) No discomfort
 - (b) Slight discomfort
 - (c) Moderate discomfort
 - (d) A lot of discomfort
 - (e) Severe discomfort
- 3 Does your voice get worse as you talk?
 - (a) Not at all – it stays the same
 - (b) Occasionally when I talk
 - (c) Often gets worse when I talk
 - (d) Often gets a lot worse when I talk
 - (e) Always gets a lot worse when I talk
- 4 Do you find it an effort to talk?
 - (a) No effort at all
 - (b) Slight effort sometimes (i.e. at the end of the day or when talking loudly)
 - (c) Quite an effort sometimes
 - (d) An effort most of the time
 - (e) A constant effort
- 5 How much are you using your voice at present?
 - (a) As much as I usually would
 - (b) A little less than I usually would
 - (c) Somewhat less than usual
 - (d) A lot less than usual
 - (e) Hardly at all
- 6 Does your voice problem stop you from doing anything that you would otherwise normally do?
 - (a) Doesn't stop me doing anything that involves me using my voice
 - (b) Stops me doing a few things that involve using my voice



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