INTERNATIONAL CO-OPERATIVE ATAXIA RATING SCALE

World Federation of Neurology

I. Posture and gait disturbances

1. Walking capacities

(Observed during a 10-meter test including a half-turn, near a wall, at about 1.5 meter.)

• 0 = Normal

• 1 = Almost normal naturally, but unable to walk with feet in tandem position

• 2 = Walking without support, but clearly abnormal and irregular

• 3 = Walking without support but with considerable staggering; difficulties in half turn

• 4 = Walking with autonomous support no longer possible; the patients uses the episodic support of the wall for a 10-meter test

• 5 = Walking only possible with one stick

• 6 = Walking only possible with two special sticks or with a stroller

• 7 = Walking only with accompanying person

• 8 = Walking impossible, even with accompanying person (wheelchair)

Score: …

2. Gait speed

(Observed in patients with preceding scores 1–3; preceding score 4 and up gives automatically score 4 in this test.)

• 0 = Normal

• 1 = Slightly reduced

• 2 = Markedly reduced

• 3 = Extremely slow

• 4 = Walking with autonomous support no longer possible

Score: …

3. Standing capacities, eyes open

(The patient is asked first to try to stay on one foot, if impossible, to stand with feet in tandem position; if impossible, to stand feet together; for the natural position, the patient is asked to find a comfortable standing position).

• 0 = Normal: able to stand on one foot more than 10 s

• 1 = Able to stand with feet together, but no longer able to stand on one foot more than 10 s

• 2 = Able to stand with feet together, but no longer able to stand with feet in tandem position

• 3 = No longer able to stand with feet together, but able to stand in natural position without support, with moderate sway

• 4 = Standing in natural position without support, with considerable sway and considerable corrections

• 5 = Unable to stand in natural position without strong support of one arm

• 6 = Unable to stand at all, even with strong support of two arms

Score: …

4. Spread of feet in natural position without support, eyes open

(The patient is asked to find a comfortable position; then the distance between medial malleoli is measured.)

• 0 = Normal (< 10 cm)

• 1 = Slightly enlarged (> 10 cm)

• 2 = Clearly enlarged (25 cm < spread < 35 cm)

• 3 = Severely enlarged (> 35 cm)

• 4 = Standing in natural position impossible

Score: …

5. Body sway with feet together, eyes open

• 0 = Normal

• 1 = Slight oscillations

• 2 = Moderate oscillations (< 10 cm at the level of head)

• 3 = Severe oscillations (> 10 cm at the level of head), threatening the upright position

• 4 = Immediate falling

Score: …

6. Body sway with feet together, eyes closed

• 0 = Normal

• 1 = Slight oscillations

• 2 = Moderate oscillations (< 10 cm at the level of head)

• 3 = Severe oscillations (> 10 cm at the level of head), threatening the upright position

• 4 = Immediate falling

Score: …

7. Quality of sitting position

(Thighs together, on a hard surface, arms folded).

• 0 = Normal

• 1 = With slight oscillations of the trunk

• 2 = With moderate oscillations of the trunk and legs

• 3 = With severe dysequilibrium

• 4 = Impossible

Score: …**Posture and gait score (static score): …/34**

II. Kinetic functions

8. Knee-tibia test (decomposition of movement and intention tremor)

(The test is performed in the supine position, but the head is tilted, so that visual control is possible. The patient is requested to raise one leg and place the heel on the knee, and then slide the heel down the anterior tibial surface of the resting leg towards the ankle. On reaching the ankle joint, the leg is again raised in the air to a height of approximately 40 cms and the action is repeated. At least 3 movements of each limb must be performed for proper assessment.)

• 0 = Normal

• 1 = Lowering of heel in continuous axis, but the movement is decomposed in several phases, without real jerks, or abnormally slow

• 2 = Lowering jerkily in the axis

• 3 = Lowering jerkily with lateral movements

• 4 = Lowering jerkily with extremely strong lateral movements or test impossible

Score right: …Score left: …

9. Action tremor in the heel-to-knee test

(Same test as preceding one: the action tremor of the heel on the knee is specifically observed when the patient holds the heel on the knee for a few seconds before sliding down the anterior tibial surface; visual control is required.)

• 0 = No trouble

• 1 = Tremor stopping immediately when the heel reaches the knee

• 2 = Tremor stopping in less than 10 seconds after reaching the knee

• 3 = Tremor continuing for more than 10 seconds after reaching the knee

• 4 = Uninterrupted tremor or test impossible

Score right: …Score left: …

10. Finger-to-nose test: decomposition and dysmetria

(The subject sits on a chair; the hand is resting on the knee before the beginning of the movement; visual control is required. Three movements of each limb must be performed for proper assessment.)

• 0 = No trouble

• 1 = Oscillating movement without decomposition of the movement

• 2 = Segmented movement in 2 phases and/or moderate dysmetria in reaching nose

• 3 = Segmented movement in more than 2 phases and/or considerable dysmetria in reaching nose

• 4 = Dysmetria preventing the patient from reaching nose

Score right: …Score left: …

11. Finger-to-nose test: intention tremor of the finger

(The studied tremor is that appearing during the ballistic phase of the movement; the patient is sitting comfortably, with his hand resting on his/her thigh; visual control is required; three movements of each limb must be performed for proper assessment.)

• 0 = No trouble

• 1 = Simple swerve of the movement

• 2 = Moderate tremor with estimated amplitude < 10 cm

• 3 = Tremor with estimated amplitude between 10 cm and 40 cm

• 4 = Severe tremor with estimated amplitude > 40 cm

Score right: …Score left: …

12. Finger-finger test (action tremor and/or instability)

(The sitting patient is asked to maintain medially his/her 2 index fingers pointing at each other for about 10 s, at a distance of about 1 cm, at the level of the thorax, under visual control.)

• 0 = Normal

• 1 = Mild instability

• 2 = Moderate oscillations of finger with estimated amplitude < 10 cm

• 3 = Considerable oscillations of finger with estimated amplitude between 10 and 40 cm

• 4 = Jerky movements > 40 cm of amplitude

Score right: …Score left: …

13. Pronation-supination alternating movements

(The subject, comfortably sitting on a chair, is asked to raise his/her forearm vertically and to make alternative movements of the hand. Each hand is moved and assessed separately.)

• 0 = Normal

• 1 = Slightly irregular and slowed

• 2 = Clearly irregular, and slowed, but without sway of the elbow

• 3 = Extremely irregular and slowed movement, with sway of the elbow

• 4 = Movement completely disorganised or impossible

Score right: …Score left: …

14. Drawing of the Archimedes' spiral on a predrawn pattern

(The subject is comfortably settled in front of a table, the sheet of paper being fixed to avoid artefacts. The subject is asked to perform the task without timing requirements. The same conditions of examination must be used at each examination: same table, same pen. The dominant hand is examined. For assessment, see joined examples at the end of this section.)

• 0 = Normal

• 1 = Impairment and decomposition, the line quitting the pattern slightly, but without hypermetric swerve

• 2 = Line completely out of the pattern with recrossings and/or hypermetric swerves

• 3 = Major disturbance due to hypermetria and decomposition

• 4 = Drawing completely disorganised or impossible

Score: …**Kinetic score (limb coordination): …/52**

III. Speech disorders

15. Dysarthria: fluency of speech

(The patient is asked to repeat several times a standard sentence, always the same, for instance: “A mischievous spectacle in Czecoslovakia”.)

• 0 = Normal

• 1 = Mild modification of fluency

• 2 = Moderate modification of fluency

• 3 = Considerably slow and dysarthric speech

• 4 = No speech

Score: …

16. Dysarthria: clarity of speech

• 0 = Normal

• 1 = Suggestion of slurring

• 2 = Definite slurring, most words understandable

• 3 = Severe slurring, speech not understandable

• 4 = No speech

Score: …**Dysarthria score: …/8**

IV. Oculomotor disorders

17. Gaze-evoked nystagmus

(The subject is asked to look laterally at the finger of the examiner: the movements assessed are mainly horizontal, but they may be oblique, rotatory, or vertical.)

• 0 = Normal

• 1 = Transient

• 2 = Persistent but moderate

• 3 = Persistent and severe

Score: …

18. Abnormalities of the ocular pursuit

(The subject is asked to follow the slow lateral movement performed by the finger of the examiner.)

• 0 = Normal

• 1 = Slightly saccadic

• 2 = Clearly saccadic

Score: …

19. Dysmetria of the saccade

(The two index fingers of the examiner are placed in each temporal visual field of the patient, whose eyes are in the primary position; the patient is then asked to look laterally at the finger, on the right and on the left; the average overshoot or undershoot of the two sides is then estimated.)

• 0 = Absent

• 1 = Bilateral clear overshoot or undershoot of the saccade

Score: …**Oculomotor movement score: …/6 TOTAL ATAXIA SCORE: …/100**