

Field Testing of an Ataxia Scoring and Staging System

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SUMMARY: *The authors present a simple system for disability scoring and functional staging of an ataxic patient, based on modifications of a previous scheme advocated by De Falco and collaborators (1979). This system was tested under field conditions in 47 ataxic subjects and found to be useful and functional.*

RÉSUMÉ: *Les auteurs présentent un système simple pour la quantification et la classification fonctionnelle d'un patient ataxique. Ce système est basé sur des modifications du schéma publié en 1979 par De Falco et collaborateurs. Notre système fut vérifié dans des conditions d'expédition sur place chez 47 patients ataxiques et trouvé utile et adéquat.*

Two of the most difficult problems facing the clinician in the field of ataxia are those of classification and the various phenotypic presentations and quantification of the signs and symptoms observed. Such grading and staging systems have been utilized with success for disorders of the basal ganglia (Hoehn and Yahr, 1967) and have permitted the evaluation of new therapeutic approaches with objective data. Unfortunately, such systems did not exist for ataxic disorders until recently. In 1979, De Falco et al, proposed a workable system which they tested in their own patients of Southern Italy. The purpose of the present study was to test the workability of this system during investigations of ataxic patients in their homes and far from the research clinic. Experience with the De Falco scheme led to modifications which were also tested and will be detailed.

SUBJECTS AND METHODS

During a two week period in the month of July 1980, on the occasion of our yearly field investigation of ataxic patients, a total of 47 subjects (26 men; 21 women) were studied by a team of two neurologists (E.P. and A.B.). The type of ataxics involved are listed in Table One. Four patients had all the symptoms of typical Friedreich's ataxia as described by Geoffroy et al (1976). Three suffered from what we have termed "Ataxia of Beauce" (Barbeau, 1980). These patients had a recessive disorder characterized by gait and truncal ataxia with pyramidal tract deficits and no signs of posterior column involvement. It will be noted that, until the biochemical marker is known, we have elected to assign geographic names to the phenotypic subtypes examined. This takes into account the genetic and geographic isolate factors often encountered. Two

patients had olivo-cerebellar atrophies (OPCA) of a recessive type, mainly characterized by speech disorders and gait ataxia (so called Matane type). Twenty patients suffered from a dominant form of olivo-ponto-cerebellar atrophy found in the Gaspé peninsula and previously reported by us (Washtiaux et al, 1978). The other 18 patients were examined in Minneapolis through the courtesy of Dr. Larry Schut, during a yearly follow up clinic of the National Ataxia Foundation. They included one female patient with the Sanger-Brown variant of OPCA (spasticity and macular degeneration), two OPCA type I (Konigsmark and Weiner, 1970) from Wyoming and 15 members of the well known Schut-Swier OPCA family (Schut 1950, Landis et al, 1974) including 5 asymptomatic at risk subjects, and 10 clear-cut ataxics.

All patients were examined at home (or at the Minneapolis Clinic) by the two neurologists working as a team. While one carried out a standard neurological examination, the other one qualified every symptom and sign on a 0 to 3 scale (0 = normal, 1 = slight; 2 = moderate and 3 = severe disability) and simultaneously with the scale reported by De Falco et al (1979). Both neurologists then decided on the functional stage to ascribe to each patient based on a scheme modified from De Falco et al (1979) and listed in Table 2.

RESULTS AND DISCUSSION

All 47 patients kindly collaborated in this detailed examination procedure. The initial evaluation of the data was based on the exact reproduction of the scheme developed by the Italian authors. This scheme lists 26 scored items. It proved to be adequate, as can be seen in the paper by Campanella et al in this issue, but incomplete and not

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TABLE 1
List of Patients Examined

Number of Type of Ataxia	Subjects Examined	Genetics	Ethnic Origin
1. Friedreich's Ataxia	4	Recessive	French canadian
2. Ataxia of Beauce	3	Recessive	French canadian
3. Sanger-Brown cerebellar atrophy	1	Dominant	Scandinavian
4. O.P.C.A. (Wyoming type)	2	Dominant	Anglosaxon
5. O.P.C.A. (Matane type)	2	Recessive	French canadian
6. O.P.C.A. (Schut-Swier type)		Dominant	Dutch
— at risk subjects	5		
— ataxic subjects	10		
7. O.P.C.A. (Gaspésie type)	20	Dominant	French canadian
TOTAL	47		

sufficiently discriminative. For example, some items are scored on a 0 - 2 scale, and others on a 0 - 4 scale, making it difficult to function in the field, without the criteria master sheet. Moreover, some items of importance in the diagnosis of certain forms of ataxia are not included, such as muscle strength, pes cavus, scoliosis, postural tremor. We were forced to add copious notes to the recording sheets to reflect the complete picture.

For all these reasons we decided to prepare a new standardized grading form (Tables 3 and 4) which permits a discriminant analysis of major sub-scores: cranial nerves, coordination, tone, reflexes, peripheral signs and muscle strength. Criteria and definitions are listed in Table 5. It should be noted that all scores are graded on a scale of 0 - 3 which is probably the one offering the most familiarity to the clinician: slight, moderate and severe disability or deviation from normal. The 47 patients in our study were thus graded and the averaged results are given in Table 6. It can be seen that there was a significant positive correlation between the total ataxia score and functional stages. A more detailed analysis of each sub-score (not reported here for lack of space but to be included in a thesis by E.P.) would reveal for example that OPCA patients have high coordination and tone scores while Friedreich's ataxia patients have very high ataxia scores coupled with maximal deviation from normal (decrease) in the tendon reflex score, severe muscle weakness scores

TABLE 2
Functional Staging of Ataxia

STAGE 0:	Normal in at-risk subject.
STAGE I:	Patient with minimal signs detected during family screening, but previously undetected and unsuspected by patient.
STAGE II:	Symptoms present, recognized by patient, but still mild. The patient is physically capable of leading an independent life, but work activities can be somewhat restricted.
STAGE III:	Symptoms are completely developed. The patient needs help or aids to move about and transfer.
STAGE IV:	Patient is confined to wheel chair.
STAGE V:	Patient is confined to bed.

(Table 7), and marked peripheral signs scores.

We feel that this scoring and staging system is an easy, workable and reproducible scheme for objectively grading the disability of an ataxic patient, even under field conditions. We plan to use this scheme in a series of pharmacologic trials to be undertaken shortly.

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TABLE 3
Ataxia Clinical Rating Scale

A. IDENTIFICATION
 Name: _____ Date exam: _____
 Address: _____ Examiner: _____
 Treating physician: _____ Chart No.: _____

B. EVOLUTION
 Present age: _____ Known heredity: _____
 Age at onset of symptoms: _____ Handedness: _____
 Sex: _____

C. DIAGRAM OF SPIRAL
 (right and left hand)

D. SUB-SCORES FUNCTIONAL STAGE	Heredity	Age Onset	Duration illness (years)	Cranial nerves	Ataxia	Tone		Re- flexes		Peri- pheral	Muscle str.	Total Score
						+/-	+/-					
MAX. 138												

E. CODING **F. FINAL DIAGNOSIS**

TABLE 4
Ataxia Clinical Rating Scale

All items on 0 - 3 scale: 0: N; 1: Mild; 2: Moderate; 3: Severe disability

Items	Score	Items	Score
A. CRANIAL NERVES		B. COORDINATION	
1. Speech		7. Normal gait	
2. Nystagmus		8. Tandem gait	
3. Eye movements		9. Romberg	
4. Fundi		10. Finger to nose — right	
5. Tongue changes		11. Finger to nose — left	
6. Cough		12. Heel to knee — right	
SUB-SCORE (C.N.)		13. Heel to knee — left	
	MAX 18	14. Spiral — right	
		15. Spiral — left	
		16. Adiadocokinesia — right	
		17. Adiadocokinesia — left	
		18. Tapping — right hand	
		19. Tapping — left hand	
		20. Postural tremor	
		SUB-SCORE (AT) MAX 42	

<p>C. TONE (deviation from N.) +/-</p> <p>21. Tone — right arm</p> <p>22. Tone — left arm</p> <p>23. Tone — right leg</p> <p>24. Tone — left leg</p> <p>25. Holmes test — right</p> <p>26. Holmes test — left</p> <hr/> <p style="text-align: right;">SUB-SCORE (T) MAX 18</p>	<p>D. REFLEXES (deviation from N.) +/-</p> <p>27. DTR — right biceps</p> <p>28. DTR — left biceps</p> <p>29. DTR — right knee</p> <p>30. DTR — left knee</p> <p>31. DTR — right achil.</p> <p>32. DTR — left achil.</p> <p>33. Babinski — right</p> <p>34. Babinski — left</p> <hr/> <p style="text-align: right;">SUB-SCORE (R) MAX 24</p>
<p>E. PERIPHERAL SIGNS</p> <p>35. Atrophy upper limbs</p> <p>36. Atrophy lower limbs</p> <p>37. Fasciculations</p> <p>38. Myoclonias</p> <p>39. Pes cavus</p> <p>40. Scoliosis</p> <p>41. Vibrations — right (secs)</p> <p>42. Vibrations — left (secs)</p> <hr/> <p style="text-align: right;">SUB-SCORE (P.S.) MAX 24</p>	<p>F. MUSCLE STRENGTH DECREASE</p> <p>43. Right arm</p> <p>44. Left arm</p> <p>45. Right leg</p> <p>46. Left leg</p> <hr/> <p style="text-align: right;">SUB-SCORE (MS) MAX 12</p>

TABLE 5

Ataxia Clinical Rating Scale Criteria and Definitions

GENERAL NOTES: Unless otherwise indicated, all items are coded on a 0 to 3 scale where
 0: normal
 1: mild disability or deviation from normal
 2: moderate disability or deviation from normal
 3: severe disability or deviation from normal

SPECIFIC CRITERIA (Number corresponds to item; items not quoted, see above):

Item	Title	Grades
2.	NYSTAGMUS	1. fine lateral nystagmus 2. clear lateral +/- upward gaze nystagmus 3. spontaneous nystagmus
3.	EYE MOVEMENTS	1. smooth but slow pursuit (viscosity) 2. saccadic or irregular pursuit 3. paralysis in one or more directions
4.	FUNDI	1. slight temporal palor 2. retinitis +/- optic atrophy 3. macular or retinal degeneration + optic atrophy
5.	TONGUE	1. fasciculations alone 2. fasciculations + slight atrophy 3. marked atrophy + limitation movements
6.	COUGH	1. hesitation in 3 consecutive coughs 2. dyspnea and arrest in consc. coughs 3. unable to do test

7, 8	NORMAL GAIT/TANDEM	1. dysbalance but gait without support 2. dysbalance but needs support 3. gait or tandem impossible
9.	ROMBERG	1. some oscillations eyes closed 2. oscillations eyes open 3. manoeuvre impossible
16, 17.	"TEST DES MARIONETTES"	
20.	POSTURAL TREMOR	1. fine hand tremor (extended arm) 2. marked hand tremor 3. hand, trunk and hand tremor
D.	TONE & REFLEXES	To be scored as deviation from normal (mild, moderate, severe) with direction indicated: increased: +; decreased: -; (absent is scored as -3; presence of clonus implies +3)
25, 26.	HOLMES TEST	0. normal braking 1. braking present but delayed 2. arms hits examiner's protecting arm 3. arm would slap face or thorax without protection
33, 34.	BABINSKI TEST	1. no response to test (indifferent) 2. fanning of toes alone 3. fanning and slow big toe retraction
37.	FASCICULATIONS	1. rare fasciculations 2. localized but frequent fasciculations 3. generalized fasciculations
38.	MYOCLONIAS 2. myoclonias bilateral, occasional	1. rare myoclonias, unilateral 3. continuous myoclonias
39.	FEET	1. high arches 2. definite pes cavus 3. pes cavus + toe deformities
40.	SCOLIOSIS	1. scoliosis 0 - 10° 2. scoliosis 10 - 30° 3. scoliosis > 30°
41, 42.	VIBRATIONS	Taken with 128 cps tuning-fork at external malleolus 0. 11 or more secs 2. 5 - 8 secs 1. 8 - 11 secs 3. < 5 secs
43, 44, 45, 46.	MUSCLE STRENGTH	1. weakness upon testing only 2. difficulty in maintaining posture or resisting a pull 3. total paralysis

TABLE 6
Ataxia Clinical Rating Results

Functional Stage	n	Mean Total Score	± S.E.M.	Range
0 (at risk)	5	4.4	1.2	0 - 7
1	7	13.0	1.2	9 - 17
2	13	41.0	1.9	31 - 54
3	11	67.9	3.8	51 - 88
4	11	84.1	4.9	54 - 107

TABLE 7
Mean Scores in Various Forms of Ataxia
(mean \pm SEM)

No. of ataxic patients	OPCA (Gaspé) 20	OPCA (Minn.) 10	Friedreich's 4	Beauce 3
mean age of onset	30.1 \pm 3.1	20.6 \pm 1.7	15.7 \pm 0.9	17.7 \pm 3.8
duration of illness (yrs)	13.6 \pm 2.1	3.2 \pm 0.8	16.0 \pm 1.6	24.0 \pm 8.5
cranial nerve defects	3.5 \pm 0.5	4.9 \pm 1.1	7.5 \pm 1.9	4.0 \pm 0.6
coordination defects	24.4 \pm 2.0	15.2 \pm 3.9	29.5 \pm 2.1	29.7 \pm 4.3
muscle tone changes	7.4 \pm 1.0	9.1 \pm 1.0	10.5 \pm 1.3	10.7 \pm 4.4
changes in reflexes	10.9 \pm 1.5	8.9 \pm 1.9	24.0 \pm 0.0	13.3 \pm 3.4
peripheral signs	3.5 \pm 0.9	0.8 \pm 0.3	11.7 \pm 0.7	7.7 \pm 5.0
muscle strength changes	2.7 \pm 0.8	1.5 \pm 0.9	8.5 \pm 1.0	4.7 \pm 2.9