Field Testing of an Ataxia Scoring and Staging System

EMMANUELLE POURCHER and ANDRE BARBEAU

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.

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 (Was-tiaux 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
The 47 patients in our study were thus evaluated 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 undertaken shortly).

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.

ACKNOWLEDGMENTS

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REFERENCES


<table>
<thead>
<tr>
<th>Number of Type of Ataxia</th>
<th>Subjects Examined</th>
<th>Genetics</th>
<th>Ethnic Origin</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Friedreich's Ataxia</td>
<td>4</td>
<td>Recessive</td>
<td>French Canadian</td>
</tr>
<tr>
<td>2. Ataxia of Beauce</td>
<td>3</td>
<td>Recessive</td>
<td>French Canadian</td>
</tr>
<tr>
<td>3. Sanger-Brown cerebellar atrophy</td>
<td>1</td>
<td>Dominant</td>
<td>Scandinavian</td>
</tr>
<tr>
<td>4. O.P.C.A. (Wyoming type)</td>
<td>2</td>
<td>Dominant</td>
<td>Anglosaxon</td>
</tr>
<tr>
<td>5. O.P.C.A. (Matane type)</td>
<td>2</td>
<td>Recessive</td>
<td>French Canadian</td>
</tr>
<tr>
<td>6. O.P.C.A. (Schut-Swier type)</td>
<td>5</td>
<td>Dominant</td>
<td>Dutch</td>
</tr>
<tr>
<td>— at risk subjects</td>
<td>10</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. O.P.C.A. (Gaspésie type)</td>
<td>20</td>
<td>Dominant</td>
<td>French Canadian</td>
</tr>
</tbody>
</table>

TOTAL 47

<table>
<thead>
<tr>
<th>Staging</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>STAGE 0</td>
<td>Normal in at-risk subject.</td>
</tr>
<tr>
<td>STAGE I</td>
<td>Patient with minimal signs detected during family screening, but previously undetected and unsuspected by patient.</td>
</tr>
<tr>
<td>STAGE II</td>
<td>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.</td>
</tr>
<tr>
<td>STAGE III</td>
<td>Symptoms are completely developed. The patient needs help or aids to move about and transfer.</td>
</tr>
<tr>
<td>STAGE IV</td>
<td>Patient is confined to wheel chair.</td>
</tr>
<tr>
<td>STAGE V</td>
<td>Patient is confined to bed.</td>
</tr>
</tbody>
</table>
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

<table>
<thead>
<tr>
<th>Heredity</th>
<th>Age Onset</th>
<th>Duration illness (years)</th>
<th>Cranial nerves</th>
<th>Ataxia</th>
<th>Tone</th>
<th>Reflexes</th>
<th>Peri­pheral</th>
<th>Muscle str.</th>
<th>Total Score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+/-</td>
<td>+/-</td>
<td></td>
<td></td>
<td>MAX 138</td>
</tr>
</tbody>
</table>

E. CODING

F. FINAL DIAGNOSIS

TABLE 4
Ataxia Clinical Rating Scale

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

A. CRANIAL NERVES
1. Speech
2. Nystagmus
3. Eye movements
4. Fundi
5. Tongue changes
6. Cough

SUB-SCORE (C.N.) MAX 18

B. COORDINATION
7. Normal gait
8. Tandem gait
9. Romberg
10. Finger to nose — right
11. Finger to nose — left
12. Heel to knee — right
13. Heel to knee — left
14. Spiral — right
15. Spiral — left
16. Adiabocokinesia — right
17. Adiabocokinesia — left
18. Tapping — right hand
19. Tapping — left hand
20. Postural tremor

SUB-SCORE (AT) MAX 42
### 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):

<table>
<thead>
<tr>
<th>Item</th>
<th>Title</th>
<th>Grades</th>
</tr>
</thead>
</table>
| 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  
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*

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

*Pourcher & Barbeau*
### TABLE 7

**Mean Scores in Various Forms of Ataxia**

*(mean ± SEM)*

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