FAP 5

SUPPLEMENTAL DATA: E - APPENDIX I (E)A-1): RATING SCALE FOR FRIEDREICH'S ATAXIA

I. FUNCTIONAL STAGING FOR ATAXIA

Increment by 0.5 may be used if the status is about the middle between two stages.

STAGE ☐☐

STAGE 0: Normal.

STAGE 1.0: Minimal signs detected by physician during screening. Can run or jump without loss of balance. No disability.

STAGE 2.0: Symptoms present, recognized by patient, but still mild. Cannot run or jump without losing balance. The patient is physically capable of leading an independent life, but daily activities may be somewhat restricted. Minimal disability.

STAGE 3.0: Symptoms are overt and significant. Requires regular or periodic holding onto wall/furniture or use of a cane for stability and walking. Mild disability. (Note: many patients postpone obtaining a cane by avoiding open spaces and walking with the aid of walls/ people etc. These patients are grades as stage 3.0)

STAGE 4.0: Walking requires a walker, Canadian crutches or two canes. Or other aids such as walking dogs. Can perform several activities of daily living. Moderate disability.

STAGE 5.0: Confined but can navigate a wheelchair. Can perform some activities of daily living that do not require standing or walking. Severe disability.

STAGE 6.0: Confined to wheelchair or bed with total dependency for all activities of daily living. Total disability.

II. ACTIVITIES OF DAILY LIVING (increments of 0.5 may be used if strongly felt that a task falls between 2 scores)

1. Speech

0 - Normal
1 - Mildly affected. No difficulty being understood.
2 - Moderately affected. Sometimes asked to repeat statements.
3 - Severely affected. Frequently asked to repeat statements.
4 - Unintelligible most of the time.

2. Swallowing

0 - Normal.
1 - Rare choking (< once a month).
2 - Frequent choking (< once a week, > once a month).
3 - Requires modified food or chokes multiple times a week. Or patient avoids certain foods.
4 - Requires NG tube or gastrostomy feedings.

3. Cutting Food and Handling Utensils

0 - Normal.
1 - Somewhat slow and clumsy, but no help needed.
2 - Clumsy and slow, but can cut most foods with some help needed. Or needs assistance when in a hurry.
3 - Food must be cut by someone, but can still feed self slowly.
4 - Needs to be fed.

4. Dressing

0 - Normal.
1 - Somewhat slow, but no help needed.
2 - Occasional assistance with buttoning, getting arms in sleeves, etc. or has to modify activity in some way (e.g. Having to sit to get dressed; use velcro for shoes, stop wearing ties, etc.).
3 - Considerable help required, but can do some things alone.
4 - Helpless.

5. Personal Hygiene

0 - Normal.
1 - Somewhat slow, but no help needed.
2 - Very slow hygienic care or has need for devices such as special grab bars, tub bench, shower chair, etc.
3 - Requires personal help with washing, brushing teeth, combing hair or using toilet.
4 - Fully dependent

6. Falling (assistive device = score 3)

0 - Normal.
1 - Rare falling (< once a month).
2 - Occasional falls (once a week to once a month).
3 - Falls multiple times a week or requires device to prevent falls.
4 - Unable to stand or walk.

7. Walking (assistive device = score 3)

0 - Normal.
1 - Mild difficulty, perception of imbalance.
2 - Moderate difficulty, but requires little or no assistance.
3 - Severe disturbance of walking, requires assistance or walking aids.
4 - Cannot walk at all even with assistance (wheelchair bound).

8. Quality of Sitting Position

0 - Normal.
1 - Slight imbalance of the trunk, but needs no back support.
2 - Unable to sit without back support.
3 - Can sit only with extensive support (Geriatric chair, posy, etc.).
4 - Unable to sit.

9. Bladder Function (if using drugs for bladder, automatic score of 3)

0 - Normal.
1 - Mild urinary hesitance, urgency or retention (< once a month).
2 - Moderate hesitance, urgency, rare retention/incontinence (> once a month, but < once a week).
3 - Frequent urinary incontinence (> once a week).
4 - Loss of bladder function requiring intermittent catheterization/indwelling catheter.

TOTAL ACTIVITIES OF DAILY LIVING SCORE:

III. NEUROLOGICAL EXAMINATION (rate each item on the basis of the patient status during examination. To the extent possible, sequential patient examinations should be carried out at the same time of the day. If the patient is taking any medication, the examination should be carried out prior to dosing, or at a fixed time following the dosing based on the maximum expected therapeutic response. Increments of 0.5 may be used if examiner feels an item falls between 2 defined severities)

A. BULBAR

1. Facial Atrophy, Fasciculation, Action Myoclonus, and Weakness:

0 - None
1 - Fasciculations or action myoclonus, but no atrophy.
2 - Atrophy present but not profound or complete.
3 - Profound atrophy and weakness.

2. Tongue Atrophy, Fasciculation, Action Myoclonus and Weakness:

0 - None.
1 - Fasciculations or action myoclonus, but no atrophy.
2 - Atrophy present but not profound or complete.
3 - Profound atrophy and weakness.

3. Cough: (Patient asked to cough forcefully 3 times)

0 - Normal.
1 - Depressed.
2 - Totally or nearly absent.

4. Spontaneous Speech (ask the patient to read or repeat the sentences "The President lives in the White House" or "The traffic is heavy today"):

0 - Normal.
1 - Mild (all or most words understandable).
2 - Moderate (most words not understandable).
3 - Severe (no or almost no useful speech).

TOTAL BULBAR SCORE:

B. UPPER LIMB COORDINATION

1. **Finger to Finger Test** (The index fingers are placed in front of each other with flexion at the elbow about 25 cm. from the sternum. Observe for 10 seconds. Score amplitude of oscillations):

	Right	Left
0 - Normal. 1 - Mild oscillations of finger (< 2 cm.). 2 - Moderate oscillations of finger (2-6 cm.). 3 - Severe oscillations of finger (> 6 cm.).	☐	☐

2. **Nose-Finger Test** (**Assess kinetic** or intention **tremor** during and towards the end of movement; examiner holds index finger at 90% reach of patient; test at least 3 nose-finger-nose trials; movement slow > 3 sec.):

	Right	Left
0 - None 1 - Mild (< 2 cm. amplitude). 2 - Moderate (2-4 cm. amplitude or persisting through movement). 3 - Severe (> 6 cm. & persisting through movement). 4 - Too poorly coordinated to perform task.	☐	☐

3. **Dysmetria** (Fast Nose-Finger) **Test : (Assess dysmetria**: The patient touches tip of examiner's finger 8 times as rapidly as possible while the examiner moves his finger and stops st different locations at about 90% reach of the patient. Assess dysmetria – i.e. inaccuracy of reaching the target- at examiner's finger):

	Right	Left
0 - None. 1 - Mild (misses 2 or fewer times). 2 - Moderate (misses 3-5 times). 3 - Severe (misses 6-8 times.). 4 - Too poorly coordinated to perform task.	☐	☐

4. **Rapid Alternating Movements of Hands** (Forearm pronation/supination 15 cm. above thigh; 10 full cycles as fast as possible; assess rate, rhythm, accuracy; practice 10 cycles before rating, if time > 7 sec. add 1 to score. Use stopwatch):

	Right	Left
0 - Normal. 1 - Mild (slightly irregular or slowed). 2 - Moderate (irregular and slowed). 3 – Too poorly coordinated to perform task.	☐	☐

5. **Finger Taps** (index fingertip-to-thumb crease; 15 reps as fast as possible; practice 15 reps once before rating; if time > 6 sec., add 1 to rating. Use stopwatch):

	Right	Left
0 - Normal. 1 - Mild (misses 1-3 times). 2 - Moderate (misses 4-9 times). 3 - Severe (misses 10-15 times). 4 - Cannot perform the task.	☐	☐

TOTAL UPPER LIMB COORDINATION SCORE ☐

C. LOWER LIMB COORDINATION

1. **Heel Along Shin Slide** (under visual control, slide heel on the contralateral tibia from the patella to the ankle up and down, 3 cycles at moderate speed, 2 sec./cycle, one at a time. May be seated with contralateral leg extended or supine but perform same way each time. Circle which: supine seated):

0 - Normal (stay on shin).
1 - Mild (abnormally slow, tremulous but contact maintained).
2 - Moderate (goes off shin a total of 3 or fewer times during 3 cycles).
3 - Severe (goes off shin 4 or more times during 3 cycles).
4 - Too poorly coordinated to attempt the task.

Right ☐ Left ☐

2. **Heel-to-Shin Tap** (patient taps heel on midpoint of contralateral shin 8 times on each side from about 6-10", one at a time. May be seated with contralateral leg extended or supine but perform the same way each time. Circle which: supine seated):

0 - Normal (stays on target).
1 - Mild (misses shin 2 or < times).
2 - Moderate (misses shin 3-5 times).
3 - Severe (misses shin > 4 times).
4 - Too poorly coordinated to perform task.

Right ☐ Left ☐

TOTAL LOWER LIMB COORDINATION SCORE ☐

D. PERIPHERAL NERVOUS SYSTEM

1. **Muscle Atrophy** (score most severe atrophy in either upper or lower limb):

0 - None.
1 - Present - mild/moderate
2 - Severe/total wasting

Right ☐ Left ☐

2. **Muscle Weakness** (Test deltoids, interossei, iliopsoas and tibialis anterior. Score most severe weakness in either upper or lower limb):

0 - Normal (5/5).
1 - Mild (movement against resistance but not full power 4/5).
2 - Moderate (movement against gravity but not with added resistance 3/5)
3 - Severe (movement of joint but not against gravity 2/5).
4 - Near paralysis (muscular activity without movement 1/5).
5 - Total paralysis (0/5).

Right ☐ Left ☐

3. **Vibratory Sense** (Educate patient regarding the sensation. Tested with 128 cps tuning fork set to near full vibration; eyes closed; test over index finger and great toe. Abnormal < 15 seconds for toes and <25 seconds for hands):

	Right	Left
Time felt for toes:	_____	_____
Time felt for fingers:	_____	_____

0 - Normal.
1 - Impaired at toes.
2 - Impaired at toes or fingers.

Right ☐ Left ☐

4. **Position Sense** (test using minimal random movement of distal interphalangeal joints of index finger and big toe)

0 - Normal.
1 - Impaired at toes/or fingers.
2 - Impaired at toes and fingers.

Right ☐ Left ☐

5. **DTR** (0-absent; 1 -hyporeflexia; 2 -normal; 3 -hyperreflexia; 4 -pathologic hyperreflexia)

Right:
BJ_____ BrJ_____ KJ_____ AJ_____

Left:
BJ_____ BrJ_____ KJ_____ AJ____

0 - No areflexia.
1 - Areflexia in either upper or lower limbs.
2 - Generalized areflexia.

Right ☐ Left ☐

TOTAL PERIPHERAL NERVOUS SYSTEM SCORE ☐

E. **UPRIGHT STABILITY** (For sitting posture patient can sit in a chair or examination table. For standing and walking assessment instruct patient to wear best walking shoes and record below if barefoot, footwear or AFOs used. Stance assessment begins with feet 20 cm apart. Place marker tapes in the exam room 20 cm apart and the insides of the feet are lined up against these. Subsequent stance tests get more difficult. For feet together the entire inside of the feet should be close together as much as possible. For tandem stance, the dominant foot is in the back and the heel of the other foot is lined with the toes of the dominant foot but not in front of the toes (because this makes it even more difficult). For one foot stance, the patient is asked to stand on dominant foot and the other leg is elevated by bringing it forward with knee extended; this gives some advantage to the patient. If a patient can stand in a particular position for 1 mintues or longer in trial 1, the trials 2 and 3 are abandoned. Otherwise each of 3 trials is timed and then averaged. Grading scores are then given as noted. Tandem walk and gait are performed in a hallway. Preferably no carpet but at least serial examinations should be on the same surface. For gait place markers 25 feet apart. Patient walks the

distance turns around and comes back and the activitiy is timed. Note if the gait was achieved with or without device and serial examinations should be done with the same device as in the first examination.

Stance and gait tests may be done barefoot if patient does have appropriate footwear, however, it should be done the same way for serial measurement.)

Circle which: Barefoot Footwear

Also, indicate if AFOs are used: Yes No

1. **Sitting Posture** (Patient seated in chair with thighs together, arms folded, back unsupported; observe for 30 sec.):

   0 - Normal.
   1 - Mild oscillations of head/trunk without touching chair back or side.
   2 - Moderate oscillations of head/trunk; needs contact with chair back or side for stability.
   3 - Severe oscillations of head/trunk; needs contact with chair back or side for stability.
   4 - Support on all 4 sides for stability.

2. **Stance feet apart– Inside of feet 20 cm apart marked on floor. Use stopwatch; 3 attempts; time in seconds):**

   Trial 1 ☐ Trial 2 ☐ Trial 3 ☐ AVG ☐

   0 - 1 minute or longer.
   1 - <1 minute, >45 sec.
   2 - <45 sec., >30 sec.
   3 - <30 sec., >15 sec.
   4 - <15 sec. or needs hands held by assistant/device.

3. **Stance - Feet Together (use stopwatch; 3 attempts; time in seconds):**

   Trial 1 ☐ Trial 2 ☐ Trial 3 ☐ AVG ☐

   0 - 1 minute or longer.
   1 - <1 minute, >45 sec.
   2 - <45 sec., >30 sec.
   3 - <30 sec., >15 sec.
   4 - <15 sec.

4. **Tandem Stance** (use stopwatch; 3 attempts, dominant foot in front; time in seconds)

   Trial 1 ☐ Trial 2 ☐ Trial 3 ☐ AVG ☐

   0 - 1 minute or longer.
   1 - <1 minute, >45 sec.
   2 - <45 sec., >30 sec.
   3 - <30 sec., >15 sec.
   4 - <15 sec.

5. **Stance on Dominant Foot** (use stopwatch; 3 attempts; time in seconds):

Trial 1 ☐ Trial 2 ☐ Trial 3 ☐ AVG ☐

0 - 1 minute or longer.
1 - <1 minute, >45 sec.
2 - <45 sec., >30 sec.
3 - <30 sec., >15 sec.
4 - <15 sec.

6. **Tandem Walk** (tandem walk 10 steps in straight line; performed in hallway with no furniture within reach of 1 m / 3 ft. and no loose carpet):

0 - Normal (able to tandem walk >8 sequential steps).
1 - Able to tandem walk in < perfect manner/can tandem walk >4 sequential steps, but <8. ☐
2 - Can tandem walk, but fewer than 4 steps before losing balance.
3 - Too poorly coordinated to attempt task.

7. **Gait** (use stopwatch; walk 8 m/25 ft. at **normal pace**. turn around using single step pivot and return to start; performed in hallway with no furniture within reach of 1 m/3 ft. and no loose carpet):

Device, if any:_____

Time in seconds:_____ ☐

0 - Normal.
1 - Mild ataxia/veering/difficulty in turning; no cane/other support needed to be safe.
2 - Walks with definite ataxia; may need intermittent support/or examiner needs to walk with patient for safety sake.
3 - Moderate ataxia/veering/difficulty in turning; walking requires cane/holding onto examiner with one hand to be safe.
4 - Severe ataxia/veering; walker or both hands of examiner needed.
5 - Cannot walk even with assistance (wheelchair bound).

TOTAL UPRIGHT STABILITY SCORE

TOTAL NEUROLOGIC EXAMINATION SCORE

IV. INSTRUMENTAL TESTING

1. **PATA Rate** (Use a tape recorder that can play at slow and fast speeds (1.2 & 2.4 cm/sec). Record at normal (2.4) speed. Use a digital stopwatch. Patient seated comfortably and instructed to repeat the syllable "pata" as quickly and distinctly as possible for 10 seconds until told to stop. Start recorder and record patient's name and date. Preset stopwatch for 10 seconds. Say "go" and as soon as patient starts speaking, start timer. Say "stop" when timer beeps at end of 10 seconds. Perform test twice and count # of "patas" for each 10 seconds, using playback at slower speed. Record number for each trial and also the average score):

Trial 1 ☐ Trial 2 ☐ AVG ☐

2. **Nine-Hole Pegboard** (Make sure the stopwatch is set to zero. Introduce this section by saying, **"Now, we're going to be measuring your arm and hand function."** If this is the first visit, as, **"Are you right- or left-handed?"** Make a note of the dominant hand for subsequent instructions. Place the 9-HPT apparatus on the table directly in front of the patient. Arrange the apparatus so that the side with the pegs is in front of the hand being tested and the side with the empty pegboard is in front of the hand not being tested. Secure with Dycem. Read the following instructions to the patient: **"On this test, I want you to pick up the pegs one at a time, using one hand only, and put them into the holes as quickly as you can in any order until all the holes are filled. Then, without pausing, remove the pegs one at a time and return them to the container as quickly as you can. We'll have you do this two (2) times with each hand. We'll start with your [DOMINANT] hand. You can hold the peg board steady with you [NON-DOMINANT] hand. If a peg falls onto the table, you retrieve it and continue with the task. If a peg falls on the floor, keep working on the task and I will retrieve it for you. See how fast you can put all the pegs in and take them out again. Are your ready? Begin."**

   Start timing as soon as the patient touches the first peg, and stop timing when the last peg hits the container. If a peg drops on the floor, the examiner will retrieve it and put it back in the peg box. However, if a peg drops onto the table, the patient is to retrieve it unless it is beyond their arm reach then you can retrieve it for them. It is possible that a peg may fall beyond the reach of the examiner therefore; we recommend that you keep a few extra pegs in hand so that testing is not interrupted. Do not put extra pegs in the testing apparatus as this may confuse the subject. Record the patient's time under "Dominant hand -- Trial 1." If the subject stops after having put all the pegs into the holes, prompt the subject to move them as well by saying, "And now remove them all." If the subject begins to remove more than one peg at a time, correct him/her by saying, "Pick up one peg at a time."

   The total time to complete the task is recorded in seconds including one decimal place rounded as needed. Round up to the next tenth if hundredth's place is > = .05, round down in hundredth's place is < 0.5.)

	RIGHT		**LEFT**
Trial 1	☐	Trial 1	☐
Trial 2	☐	Trial 2	☐
AVG	☐	AVG	☐