

FA - COMS

NEUROLOGICAL EXAMINATION

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D. If the subject could not complete this test indicate why:

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- 1 = Unable to complete trial due to physical limitations – not related to FA.
- 3 = Subject was too fatigued to complete trial.
- 4 = Subject refused to complete trial.

NEUROLOGICAL EXAMINATION (rate each item on the basis of the subject status during examination. To the extent possible, sequential subject examinations should be carried out at the same time of the day. Increments of 0.5 may be used if examiner feels an item falls between 2 defined severities).

A. BULBAR

Most subjects with FA do not have significant facial or tongue atrophy. If mild facial or tongue atrophy is noted score as per instructions. Speech and Cough assessment is self-explanatory.

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

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- 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:**

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- 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: (Subject asked to cough forcefully 3 times)**

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

4. **Speech (ask the subject to read or repeat the sentences A “The President lives in the White House.” and B “The traffic is heavy today.”):**

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

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B. UPPER LIMB COORDINATION

Upper limb coordination: Most of the items are self-explanatory. For items 3 through 5, ask the subject to count as they do the task. Example: "Move your hand back and forth 10 times as fast as you can. Please count each time to yourself". You can time the activity with either a watch or a stopwatch.

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):**
- 1a. Right .
- 1b. Left .
- 0 = Normal.
1 = Mild oscillations of finger (less than 2 cm.).
2 = Moderate oscillations of finger (2-6 cm.).
3 = Severe oscillations of finger (greater than 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 subject; test at least 3 nose-finger-nose trials; movement slow greater than 3 sec.):**
- 2a. Right .
- 2b. Left .
- 0 = None.
1 = Mild (less than 2 cm. amplitude).
2 = Moderate (2-6 cm. amplitude or persisting through movement).
3 = Severe (greater than 6 cm. & persisting through movement).
4 = Too poorly coordinated to perform task.
3. **Dysmetria Test : The subject touches tip of examiner's finger then subject's chin 8 times as rapidly as possible while the examiner moves his finger to four corners of a one foot square and at about 90% reach of the subject. Assess dysmetria – (i.e. inaccuracy of reaching the target- tip of examiner's finger):**
- 3a. Right .
- 3b. 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 (Subject should be seated. 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 greater than 7 sec. add .5 to score. Use stopwatch):**
- 4a. Right .
- 4b. Left .
- 0 = Normal.
1 = Mild (slightly irregular or slowed).
2 = Moderate (irregular and slowed).
3 = Too poorly coordinated to perform task.

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B. UPPER LIMB COORDINATION (CONT)

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

5a. Right

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5b. Left

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- 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.

C. LOWER LIMB COORDINATION

Lower limb coordination: the items are self-explanatory. The heel shin slide is scored 1 if there is an abnormality but contact is steady along the top of the shin. If the heel starts going off the shin to one or other side score 2 or 3 as noted. For heel to shin tap instruct the subject to count 8 taps with heel raised about 8" each time. It is preferable to do this section with subject seated. If this is not followed for a particular subject, it should be done in the same position each time.

1. **Heel Along Shin Slide (Perform while seated, under visual control, slide heel on the contralateral tibia from the patella to the ankle up and down with contralateral leg extended, 3 cycles at moderate speed, one leg at a time):**

1a. Right

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1b. Left

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- 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 perform task.

2. **Heel-to-Shin Tap (Subject taps heel on midpoint of contralateral shin 8 times on each side from about 6-10", one at a time. Perform seated with contralateral leg extended):**

2a. Right

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2b. Left

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

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D. PERIPHERAL NERVOUS SYSTEM

Peripheral nervous system: these items are self-explanatory. Check deltoids and intrinsic hand muscles in the upper limbs; iliopsoas and tibialis anterior in the lower limbs. Atrophy and weakness are scored on the basis of the worst muscle in this group. One does not have to do extensive muscle testing. Vibration sense is recorded as noted in seconds and then given a score depending on the extent of impairment. DTR are recorded in the given space as noted and then any hypo/areflexia is given a numerical score as noted.

1. **Muscle Atrophy (score most severe atrophy in either upper or lower limb):** 1a. Right
 0 = None 1b. Left
 1 = Present - mild/moderate
 2 = Severe/total wasting
 1c. If question 1a or 1b is either 1 or 2 indicate location of atrophy:

2. **Muscle Weakness (Test deltoids, interossei, iliopsoas and tibialis anterior. Score most severe weakness in either upper or lower limb):** 2a. Right
 0 = Normal (5/5). 2b. Left
 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).

3. **Vibratory Sense** (Educate subject regarding the sensation at the elbow. Tested with 128 cps tuning fork set to near full vibration; eyes closed; test over index finger and top of great toe (most distal joint not nail). Abnormal less than 15 seconds for toes and less than 25 seconds for hands):

- 3a. Time felt for *toes* (Right) 3a. Right

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 3b. Time felt for *toes* (Left) 3b. Left

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 3c. Time felt for *fingers* (Right) 3c. Right

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 3d. Time felt for *fingers* (Left) 3d. Left

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 3e. 0 = Normal. 3e.1 Right
 1 = Impaired at toes or fingers. 3e.2 Left
 2 = Impaired at toes and fingers.

4. **Position Sense (test using minimal random movement of distal interphalangeal joints of index finger and big toe)**
 0 = Normal. 4a. Right
 1 = Impaired at toes or fingers. 4b. Left
 2 = Impaired at toes and fingers.

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5. **DTR** (0 = absent, 1 = hyporeflexia, 2 = normal, 3 = hyperreflexia, 4 = pathologic hyperreflexia)
- 5a. Right 5a.1 BJ 5a.2 BrJ 5a.3 KJ 5a.4 AJ
- 5b. Left 5b.1 BJ 5b.2 BrJ 5b.3 KJ 5b.4 AJ
- 5c. **DTR**
- 0 = No areflexia. 5c.1 Right
- 1 = Areflexia or mild hyperreflexia in either upper or lower limbs.
- 2 = Generalized areflexia or pathologic hyperreflexia. 5c.2 Left

E. UPRIGHT STABILITY

*Upright stability: For sitting posture subject can sit in a chair or examination table. For standing and walking assessment instruct subject to wear best walking shoes and record below if barefoot, footwear or AFOs [plastic brace] 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 subject 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 subject. If a subject can stand in a particular position for 1 minute or longer in trial 1 for tests 2a, 2b, 3a, 3b, 4 and 5, then 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. Subject walks the distance turns around and comes back and the activity is not 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.*

Ea. Is subject: (1 = barefoot, 2 = footwear) Ea.

Eb. Indicate if AFOs [plastic brace] are used: (0 = No, 1 = Yes) Eb.

Ec. Test performed on Carpet? (0 = No, 1 = Yes) Ec.

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

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E. UPRIGHT STABILITY (CONT)

- 2a. Stance feet apart – Inside of feet 20 cm apart marked on floor. Use stopwatch; 3 attempts; time in seconds. If greater than 60 seconds on trial 1 stop, if less than 60 seconds do all three trials):
- Length of time:**
- 0 = 1 minute or longer
 - 1 = Less than 1 minute, greater than 45 seconds
 - 2 = Less than 45 seconds, greater than 30 seconds
 - 3 = Less than 30 seconds, greater than 15 seconds
 - 4 = Less than 15 sec. or needs hands held by assistant/device
- 2a.1 Trial One
- 2a.2 Trial Two
- 2a.3 Trial Three
-
- 2b. Same as above but with eyes closed.
- Length of time:**
- 0 = 1 minute or longer
 - 1 = Less than 1 minute, greater than 45 seconds
 - 2 = Less than 45 seconds, greater than 30 seconds
 - 3 = Less than 30 seconds, greater than 15 seconds
 - 4 = Less than 15 sec. or needs hands held by assistant/device
- 2b.1 Trial One
- 2b.2 Trial Two
- 2b.3 Trial Three
-
- 3a. Stance – Feet Together (use stopwatch; 3 attempts; time in seconds):
- Length of time:**
- 0 = 1 minute or longer
 - 1 = Less than 1 minute, greater than 45 seconds
 - 2 = Less than 45 seconds, greater than 30 seconds
 - 3 = Less than 30 seconds, greater than 15 seconds
 - 4 = Less than 15 sec. or needs hands held by assistant/device
- 3a.1 Trial One
- 3a.2 Trial Two
- 3a.3 Trial Three
-
- 3b. Same as above but with eyes closed.
- Length of time:**
- 0 = 1 minute or longer
 - 1 = Less than 1 minute, greater than 45 seconds
 - 2 = Less than 45 seconds, greater than 30 seconds
 - 3 = Less than 30 seconds, greater than 15 seconds
 - 4 = Less than 15 sec. or needs hands held by assistant/device
- 3b.1 Trial One
- 3b.2 Trial Two
- 3b.3 Trial Three
-
4. Tandem Stance (dominant foot in front; front foot lined up with great toe of the back foot)
- Length of time:**
- 0 = 1 minute or longer
 - 1 = Less than 1 minute, greater than 45 seconds
 - 2 = Less than 45 seconds, greater than 30 seconds
 - 3 = Less than 30 seconds, greater than 15 seconds
 - 4 = Less than 15 sec. or needs hands held by assistant/device
- 4.1 Trial One
- 4.2 Trial Two
- 4.2.1 Trial Three

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5. Stance on Dominant Foot (Elevate leg straight out in front, use stopwatch; 3 attempts; time in seconds): 5.1 Trial One
Length of time: 5.2 Trial Two
 0 = 1 minute or longer 5.3 Trial Three
 1 = Less than 1 minute, greater than 45 seconds
 2 = Less than 45 seconds, greater than 30 seconds
 3 = Less than 30 seconds, greater than 15 seconds
 4 = Less than 15 seconds or needs hands held by assistant/device
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): 6.
 0 = Normal (able to tandem walk greater than 8 sequential steps).
 1 = Able to tandem walk in less than perfect manner/can tandem walk greater than 4 sequential steps, but less than 8.
 2 = Can tandem walk, but fewer than 4 steps before losing balance.
 3 = Too poorly coordinated to attempt task.
7. Gait (Observe subject walk at normal pace with assistive device in one direction, turn around and return to start; performed in hallway with no furniture within reach of 1 m / 3 ft. and no loose carpet): 7.
 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 subject 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).