

REV 4/10 replaces 11/09  
 MACS NEUROPSYCHOLOGICAL STUDY PHASE 2  
 NEUROLOGICAL ASSESSMENT

Form  Center

Examiner use only (not for coding)	
Age	<input type="text"/> <input type="text"/>
Handedness	<input type="text"/>
Comments/Actions:	_____
	_____

MACS STUDY ID #        
 MACS VISIT #     
 NP PHASE 2 VISIT    
 EXAM DATE   /   /    
 M M D D Y Y

REASON FOR REFERRAL   
 (1 - standard routine; 2 - control; 3 - "clinical indications";  
 4 - other)

**Q1. LIFETIME NEUROLOGICAL DISEASE:** (1 = no; 2 = yes; 9 = unknown/missing)

- |  |                      |                            |                      |
|--|----------------------|----------------------------|----------------------|
| a. Multiple sclerosis                    | <input type="text"/> | d. Seizures                | <input type="text"/> |
| b. Head trauma <u>with LOC &gt; 1 hr</u> | <input type="text"/> | e. Peripheral nerve damage | <input type="text"/> |
| c. Stroke                                | <input type="text"/> | f. Other                   | <input type="text"/> |
|  |                      | g. Slipped disc            | <input type="text"/> |

(Previous Rx, evaluation, treating MD) \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**Q2. LIFETIME SYSTEMIC DISEASE:** (1 = no; 2 = yes; 9 = unknown/missing)

- |   |                      |                                      |                      |
|---|----------------------|--------------------------------------|----------------------|
| a. Diabetes mellitus  | <input type="text"/> | d. Hypertension                      | <input type="text"/> |
| b. Connective tissue disorder<br>eg rheumatoid arthritis or lupus | <input type="text"/> | e. Remote heavy etoh<br>> 6 mos. ago | <input type="text"/> |
| c. Solvent/heavy metal exposure                                   | <input type="text"/> | f. Other lifetime illness            | <input type="text"/> |
|   |                      | g. Heavy ETOH use w/in 6 mo          | <input type="text"/> |

(Previous Rx, evaluation, treating MD) \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**Q3. ALCOHOL USE, LAST 24 HOURS:**   
 (Where one drink is equal to: 1-12 oz beer, or 1-4 oz glass of wine, or 1 oz of hard liquor)  
 (1 = none; 2 = 1 drink; 3 = 2-3drinks; 4 = 4-6 drinks; 5 = ≥ 7 drinks; 9 = unknown/missing)

Comment: \_\_\_\_\_

**Q4. NON PRESCRIPTION DRUG USE, LAST 24 HOURS**   
 (1 = none; 2 = marijuana/hashish; 3 = cocaine; 4 = opiates; 5 = uppers/downers; 6 = polysubstance;  
 7 other drugs; 9 = unknown/missing)

Comment: \_\_\_\_\_

**Q5. WORK STATUS** (Specify type of work or school in last 6 mo.)   
 (1 = stable; 2 = change in work status because of health; 3 = change for other reasons;  
 4 = change in work status > 6 mos because of health; 9 = unknown/missing)

Type of Work: \_\_\_\_\_

INSTRUCTIONS: The next set of Questions Q6-22 are hierarchical, i.e., within each question there are different sub-questions, representing increasing levels of symptomatology. The questions are designed to be asked by the Neurological Examiner, but can be asked by a separate interviewer (e.g. LA). If so, indicate separate examiner code on Page 5. Start each question with the conversational instruction using the written phrase exactly (BOXED), then ask the first sub-question (numbered as 2).

If NO: stop at that part, score as 1 (normal) and skip to next question.

If YES: continue asking successive sub-questions, ie, 3,4,5, until NO is response, score the number corresponding to highest YES, and go to next question.

Example: Question 6 in the Cognitive Function Section:

1. Start with introduction - "I'M GOING TO ASK YOU ABOUT.."
2. Ask first level of question - Do you have occasional difficulty with concentration?
3. If NO, score Q6 as 1 (normal), skip to Question Q7.
4. If YES, move to next sub-question - DO you frequently lose your concentration?

Remember: You are trying to elicit CHANGES during the past 6 months.

**Q6. CONCENTRATION  
SPEED OF THOUGHT**

**I'M GOING TO ASK YOU ABOUT NEW PROBLEMS WITH CONCENTRATION OR THINKING WITHIN THE LAST 6 MONTHS.**

- 2 - Do you have occasional difficulty with concentration?
- 3 - Do you frequently lose your concentration in conversation? Do you need help with tasks like balancing a check book?
- 4 - Are you unable to follow normal conversation? **(Examiner: score this 4 if unable to answer.)**

Comments: \_\_\_\_\_

**Q7. READING  
OR TV**

**I'M GOING TO ASK YOU ABOUT NEW DIFFICULTY WITH READING OR FOLLOWING PROGRAMS ON TV WITHIN THE LAST 6 MONTHS.**

- 2 - Do you have some trouble following programs on TV? Do you occasionally lose your place when reading?
- 3 - Have you cut down these activities because of difficulty understanding?
- 4 - Are you unable to follow a TV show or read because it doesn't make sense?

Comments: \_\_\_\_\_

**Q8. MEMORY**

**I'M GOING TO ASK YOU ABOUT NEW CHANGES IN YOUR MEMORY WITHIN THE LAST 6 MONTHS.**

- 2 - Do you have occasional problems with remembering appointments or where you put things?
- 3 - Do you frequently forget appointments or what to do in the middle of a task?
- 4 - Are you constantly confused about the time, date, or where you are?  
**(Examiner: score this 4 if participant is disoriented.)**

Comments: \_\_\_\_\_

**Q9. SPEECH**

**I'M GOING TO ASK ABOUT ANY NEW DIFFICULTIES WITH SPEAKING WITHIN THE LAST 6 MONTHS.**

- 2 - Do you have to take extra care to choose the right words occasionally?
- 3 - Do you frequently have difficulty finding words?
- 4 - Do you find it difficult to produce more than brief phrases or words?  
**(Examiner: score this 4 if participant is mute.)**

Comments: \_\_\_\_\_

**Q10. MOOD**

**I'M GOING TO ASK YOU ABOUT NEW CHANGES IN YOUR MOOD IN THE LAST 6 MONTHS.**

- 2 - Have you been feeling somewhat depressed lately?
- 3 - Has this interfered with your functioning at home or work?
- 4 - Have you needed medication or hospitalization?

Comments: \_\_\_\_\_

Q13. **GAIT**

**I'M GOING TO ASK ABOUT NEW DIFFICULTIES WITH WALKING WITHIN THE LAST 6 MONTHS.**

- 2 - Do you feel unsteady, slow, or weak when you walk?
- 3 - Are you so weak or unsteady that you require a cane?
- 4 - Are you so weak or unsteady that you require a walker?
- 5 - Are you unable to walk even with support?

Comments: \_\_\_\_\_

Q14. **COORDINATION**

**I'M GOING TO ASK ABOUT NEW CLUMSINESS IN YOUR HANDS WITHIN THE LAST 6 MONTHS.**

- 2 - Are you slower or clumsier but you can still finish everyday activities (eg. eating, buttons)?
- 3 - Do you have difficulty with everyday activities because of clumsiness?
- 4 - Do you need help with everyday activities because of clumsiness?

Comments: \_\_\_\_\_

Q15. **INVOLUNTARY MOVEMENT**

**I'M GOING TO ASK ABOUT NEW INVOLUNTARY MOVEMENTS LIKE SHAKING, TWITCHING, OR JERKING WITHIN THE LAST 6 MONTHS.**

- 2 - Have you developed any tremor or shaking in your limbs, head or trunk?
- 3 - Does it interfere with everyday activity (eg. writing)?
- 4 - Is it so severe that you cannot perform everyday activities?

Comments: \_\_\_\_\_

Q17. **SYNCOPE/ SEIZURES**

**I'M GOING TO ASK ABOUT NEW BLACKOUTS, SEIZURES, OR CONVULSIONS IN THE LAST 6 MONTHS.**

- 2 - Have you had blackouts (no seizures)?
- 3 - Have you had any seizures or convulsions?

Comments: (description and frequency) \_\_\_\_\_

Q18. **PARESTHESIAS/ DYSESTHESIAS**

**I'M GOING TO ASK YOU ABOUT NEW SENSATIONS IN YOUR ARMS AND LEGS IN THE LAST 6 MONTHS.**

- 2 - Do you have tingling or burning sensations intermittently?
- 3 - Are these feelings continuous but don't interfere with everyday activities?
- 4 - Are these feelings uncomfortable enough to limit your daily activities?

Comments: (distribution/description) \_\_\_\_\_

Q19. **LOSS OF SENSATION**

**I'M GOING TO ASK YOU ABOUT NEW NUMBNESS OR LOSS OF SENSATION IN YOUR ARMS AND LEGS IN THE LAST 6 MONTHS.**

- 2 - Have you had intermittent or fleeting loss of sensation?
- 3 - Have you noticed this continuously, but it doesn't limit everyday activities?
- 4 - Has it been severe enough to limit your daily activities?

Comments: (distribution) \_\_\_\_\_

Q20. **MUSCLE WEAKNESS**

**I'M GOING TO ASK YOU ABOUT ANY NEW WEAKNESS OF YOUR ARMS OR LEGS IN THE LAST 6 MONTHS.**

- 2 - Have you had intermittent or fleeting weakness?
  - 3 - Has it been continuous but not severe enough to limit everyday activities?
  - 4 - Has it been severe enough to limit your daily activities?
- Comments: (distribution)

Q20a. **MYALGIAS**

**I'M GOING TO ASK YOU ABOUT ANY NEW MUSCLE PAINS OR ACHING IN THE LAST 6 MONTHS.**

- 2 - Have you had intermittent or fleeting muscles aches?
  - 3 - Have you had frequent or constant aching for which you've needed medications?
  - 4 - Have you been limited in daily activities by severe muscle aching?
- Comments: (distribution)

Q21. **VISUAL**

**I'M GOING TO ASK YOU ABOUT ANY NEW PROBLEMS WITH YOUR VISION IN THE LAST 6 MONTHS.**

- 2 - Have you noticed any change of vision persisting for more than a few minutes (This could be blurring or double vision)?
  - 3 - Has this worsened to the point where it limits your daily activities?
- Comments: (ophthalmologist)

Q22. **HEADACHE**

**I'M GOING TO ASK YOU ABOUT NEW OR UNUSUAL HEADACHES IN THE LAST 6 MONTHS.**

- 2 - Have you developed more frequent or severe headaches than you are used to getting?
  - 3 - Have any of the headaches been so bad that you've had to quit work?
- Comments: (record if any nausea, vomiting, or neck stiffness)

Q23. **PHYSICIAN VISIT**

HAVE YOU CONSULTED A PHYSICIAN FOR ANY OF THE SYMPTOMS THAT WE'VE ASKED ABOUT (Q6-22)? (Record 1 for NO, 2 for YES and indicate below which symptoms)  
Comments: (Physician, address,, date, symptom, action)

Q23a. **EXAMINER CODE** (separate code ONLY if symptoms completed by examiner other than neurological examiner)

Q23b. **NEUROLOGICAL SYMPTOMS SUMMARY**

Instructions: 1 = Does not meet criteria for remaining in Phase 2. If any 3 questions (Q6-Q22) have scores  $\geq 3$ , code 2 and keep participant in Phase 2.

The neurological examination should be performed by someone with neurological training. Follow the format below and note instructions for testing. When scoring, if an abnormality is present, but you can't be sure if its new or old, record as NEW (ie 3) and write in comment section. An "old" abnormality is something that the patient knows has been present since birth, for several years, or is clearly related to trauma or surgery. If you're unsure whether the examination is abnormal, record as 2.

**Q24. RESPONSE TO COMMANDS** |\_|

1 - normal; 2 - mild slowing; 3 - moderate slowing; 4 - severe psychomotor retardation; 5 - agitated or anxious; 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing  
Comments:

**MENTAL STATUS**

**Q25. STEP COMMANDS** |\_|

COMMAND: "Take your left thumb, touch the tip of your nose, tip of your chin, right ear."  
THEN, "With your right index finger, touch your right cheek, forehead, and left shoulder."  
THEN, "With your right thumb, touch your left ear, chin, and right shoulder."

1 - normal; 2 - needs instructions repeated but performs correctly; 3 - one or more errors; 4 - cannot perform; 8 - neurologic abnormality is old, eg, right-left confusion since childhood; 9 - unknown/missing  
Comments: \_\_\_\_\_

**Q25a. REGISTRATION** |\_|

Name 3 objects: "orange, pony, quarter." Then ask the participant to repeat all three after you have said them (1 - 3 points, score 1 for each correct).

**Q25b. TIME** |\_|

Year, season, date, day, month (1 - 5 points; score 1 for each correct)

**Q25c. PLACE** |\_|

State, county or borough, town, hospital, floor (1 - 5 points; score 1 for each correct)

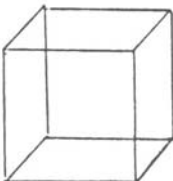
**Q25d. ATTENTION AND CONCENTRATION** |\_|

Spell "world", "horse", "black", or "bread" backwards (1 - 5 points; score number of letters correct before first error).

**Q25e. RECALL** |\_|

Ask for recall of the 3 objects repeated above (Q25a). (1 - 3 points; score 1 for each correct) Allow 3 minutes between registration and recall.

**Q25f. COPY A DESIGN** (score 1 point if accurate) |\_|



Copy here

**Q25g. TOTAL SCORE OF MENTAL STATUS** (Sum 25a-25f; maximum = 22) |\_|\_|

**Q26. FUNDUS**

1 - normal; 2 - cotton-wools spots; 3 - hemorrhage; 4 - papilledema; 5 - other or mixed;  
 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing   
 Comments: \_\_\_\_\_

**Q27. PUPILS**

Assess size, shape, symmetry, and speed of response

1 - normal; 2 - slightly sluggish; 3 - abnormal, eg. dilated, AR pupils, light-near dissociation;  
 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing   
 Comments: \_\_\_\_\_

**Q28. OCULAR MOTILITY**

Smooth pursuit: test with slow moving target, > 15" from face.

1 - normal; 2 - frequent corrections; 3 - cannot pursue or has sustained nystagmus; 8 - neurologic abnormality is old, eg, congenital nystagmus or drug effects; 9 - unknown/missing   
 Comments: \_\_\_\_\_

**Q29. SACCADES**

Test eye movement from primary position to target in all fields of gaze.

1 - normal; 2 - mildly abnormal, eg. slightly slowed; 3 - dysmetric, dysconjugate, or paretic;  
 8 - neurologic abnormality is old, congenital, post-traumatic, or drug effects; 9 - unknown/missing   
 Comments: \_\_\_\_\_

**Q30. FACIAL STRENGTH**

1 - normal; 2 - mild asymmetry or equivocal weakness; 3 - definite paresis/palsy;  
 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing   
 Comments: (pattern/side) \_\_\_\_\_

**Q31. FACIAL EXPRESSION**

1 - normal; 2 - slightly decreased blink rate and hypomotility; 3 - mask-like; 8 -neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/ missing   
 Comments: \_\_\_\_\_

**Q32. OTHER CRANIAL NERVES**

Assess speech patterns, tongue movements, palate movement, hearing.

1 normal; 2 - mild abnormality, eg. slight slurring or facial numbness; 3 - moderate/severe abnormality, eg. severe dysarthria or absent corneal reflex; 8 - neurologic abnormality is old, congenital, or post-traumatic, eg lifelong speech difficulty or deafness; 9 - unknown/missing   
 Comments: \_\_\_\_\_

**MOTOR**

Test following muscles: hand intrinsics, finger grip, wrist extensors, triceps, deltoids, foot intrinsics, toe extensors, ankle dorsiflexors, hip flexors and extensors: NOTE SCORING SYSTEM IS REVERSE OF MRC GRADE.

**Q33. STRENGTH**

1 - normal; 2 - mild, vs resistance (MRC grade 4/5) or pronator drift only; 3 - moderate, vs gravity (MRC grade 3/5); 4 - severe, (MRC grade <2/5); 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/ missing

	RU	RL	LU	LL
Proximal	a.	b.	c.	d.
Distal	e.	f.	g.	h.

Comments: (pattern/distribution)

Q34. **TONE**

Assess tone in arms and legs with patient relaxed.

1 - normal; 2 - mild hypertonia, eg. slight resistance to passive movement; 3 - moderate/severe hypertonia, eg. definite spastic catch or lead-pipe rigidity; 4 - hypotonia; 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing

RU	RL	LU	LL
a.	b.	c.	d.

Comments: (pattern/distribution) \_\_\_\_\_

34a. **TREMOR**

1 - no tremor; 2 - fine, fast "physiologic tremor;" 3 - severe, function-limiting tremor; 9 - unknown/missing

Comments: \_\_\_\_\_

Q35. **BULK**

1 normal; 2 - diffuse wasting; 3 - focal atrophy; 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing

Comments: \_\_\_\_\_

Q36. **COORDINATION AND RAM - LIMB**

Use finger-nose, heel-knee-shin, bicycling with legs, hand tap, finger-thumb, hand flip. Distinguish slowing (extrapyramidal) from inaccuracy or dysmetria (cerebellar).

1 normal; 2 - mild slowness/clumsiness - compared to examiner; 3 - moderate slowness/clumsiness - notably slow and/or dysmetric; 4 - severe, unable to perform; 5 - slowing alone with normal accuracy; 6 - weakness precludes testing; 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing

Comments: \_\_\_\_\_

Q37. **COORDINATION GAIT**

Use tandem, rapid walk with turns. Try knee bend or chair-rising to assess proximal weakness; heel-rocking for TIB ANT weakness.

1 normal; 2 - gait disturbance, evident only on rapid turns or tandem; 3 - clear difficulty with walking; 4 - severe gait disturbance, requires assistance to walk; 5 - non-ambulatory; 8 - neurologic abnormality is old, congenital, or post-traumatic; 9 - unknown/missing

Comments: \_\_\_\_\_

Q38. **TIMED GAIT**

Subject walks a 10 yd distance, turns and returns. Instruct: "Walk as fast as you can." Repeat for 2 additional trials. Score is average of 3 trials in seconds. Round up/down to nearest integer. Score 000- cannot perform; 999 - unknown/missing.

Trial 1						.
Trial 2						.
Trial 3						.
Average (secs)						.

**Q39. REFLEX GRADING**

**NOTE CHANGE IN SCORING**

0 - absent; 1 - hypoactive; 2 normal DTR's; 3 - hyperactive DTR's, eg with prominent spread;  
 4 - clonus; 9 - unknown/missing

	biceps	triceps	knee	ankle
Right	a.	b.	c.	d.
Left	e.	f.	g.	h.

**Q40. RELEASE REFLEXES**

**NOTE CHANGE IN SCORING.**

0 - absent/normal; 2 - present/abnormal; 9 - unknown/missing

	Plantars	Grasp	Snout	Jaw
Right	a.	c.	e.	f.
Left	b.	d.		

Test SNOUT by tapping with reflex hammer on upper lid with eyes lightly closed - look for pouting of lips.  
 Test JAW by tapping down on jaw with mouth held open and relaxed - look for contraction of masseters.

**Q41. SENSATION**

**A. VIBRATION**

Use tuning fork applied to big toe joint and compare duration of reported "buzzing" compared to examiner's finger under toe. If abnormal, test proprioception/sensory level, Tinel's, Rombergs.

1 - normal; 2 - mild loss, eg. vibration stops < 10 seconds before examiner; 5 - vibration stops > 10 seconds before examiner; 3 - cannot feel vibration in toes; 4 - severe, cannot feel vibration in toes and ankles; 9 - unknown/missing

Comments: \_\_\_\_\_

**B. PIN SENSATION**

Test using broken cotton wool swabstick. Compared sharp end to dull end and hands to feet.

1 - normal; 2 - mild distal loss or hemisensory difference; 3 - hyperalgesia distally, ie, patient reports that stick is more painful in feet than shins; 4 - definite distal gradient, "stocking glove" pattern, or sensory level; 5 - mixed/combination; 9 - unknown/missing

Comments: \_\_\_\_\_

**STEREOGNOSIS**

Use 2 test objects in each hand with eyes closed: eg: coin, key, paper clip.

1 - normal, recognizes both objects; 2 - recognizes 1 object; 3 - recognizes neither object; 9 - unknown

Comments: \_\_\_\_\_



**NEUROLOGIC SUMMARY**

Summary should be coded for highest level of impairment. If unsure whether a definite examination abnormality is new or old/congenital/post-traumatic, code as 3. If unsure which system is abnormal, eg whether LE hyperreflexia is "spinal cord" or "CNS diffuse," mark each system.

**Q42. PERIPHERAL NERVES**

1 - normal; 2 - equivocal/mild abnormalities, e.g. a) mild vibration loss and diminished AJ or b) mild vibration and other mild sensory modality; 3 - abnormal, definite peripheral neuropathy with absent AJ or moderate/severe vibration loss; 8 - abnormal, unrelated or old, e.g. longstanding sensory deficit s/p disc; 9 - unknown/missing

Comments: \_\_\_\_\_

**Q43. CNS DIFFUSE**

1 - normal; 2 - equivocal/mild abnormalities, e.g. diffuse hyperreflexia OR slight slowing of RAM's OR release signs; 3 - definitely abnormal, e.g. a) increased tone UE's and LE's AND hyperreflexia; OR b) slowed RAM's AND hyperreflexia; 8 - abnormal, unrelated or old, e.g. drug effects, s/p trauma; 9 - unknown/missing

Comments: \_\_\_\_\_

Q43a. Use this item for participants in whom you suspect **HIV-RELATED DEMENTIA** based on diffuse CNS signs (Q43) and/or cognitive complaints (Q6-9). Use coding from next page to grade the dementia. Grade myelopathy separately. If CNS impairment is clearly unrelated to HIV, eg post-traumatic, code as 8; 9 = unknown/missing. If only abnormalities are peripheral nerve, spinal, or muscle, code Q43a as 0. •

**Q44. CNS FOCAL**

1 - normal; 2 - equivocal/mild abnormalities, e.g. a) mild focal weakness or hyperreflexia or pronator drift OR eg b); isolated brainstem, eye movement, or pupillary abnormalities; 3 - abnormal, e.g. hemiparesis, cerebellar ataxia, aphasia; 8 - abnormal, unrelated or old, e.g. previous trauma/surgery/CVA; 9 - unknown/missing

Comments: \_\_\_\_\_

**Q45. SPINAL CORD**

1 - normal; 2 - equivocal/mild abnormalities, e.g. slightly increased tone in LE or LE hyperreflexia, but UE's normal; 3 - abnormal, e.g. spastic paraparesis with increased LE tone, reflexes and weakness or sensory level; 8 - abnormal, unrelated or old, e.g. previous trauma, s/p disc; 9 - unknown/missing

Comments: \_\_\_\_\_

Q45a. Use this item for participants in whom you suspect **HIV-RELATED MYELOPATHY** based on diffuse spinal cord signs (Q45). If myelopathy is clearly unrelated to HIV, eg post-traumatic, code as 8; 9 = unknown/missing. Use coding from next page to grade the myelopathy. Grade dementia separately.

**Q46. MUSCLE**

1 - normal; 2 - equivocal/mild abnormalities, e.g. slight proximal weakness or diffuse wasting; 3 - abnormal, e.g. proximal myopathy; 8 - abnormal, unrelated or old; 9 - unknown/missing

Comments: \_\_\_\_\_

**CODING FOR Q43A, DEMENTIA SEVERITY.**

Stage 0 (normal): Normal mental and motor function. Neurological signs are within the normal age-appropriate spectrum.

Stage 0.5 (equivocal or subclinical): Absent, minimal or equivocal symptoms without impairment of work or capacity to perform activities of daily living (ADL). Exam may be normal or mildly abnormal signs may include reflex changes (eg, generalized increase in deep tendon reflexes with active jaw jerk, snout or glabellar sign) or mildly slowed ocular movements, but without clear slowing of extremity movements or loss of their dexterity or strength.

Stage 1 (mild): Able to perform all but the more demanding aspects of work or ADL but with unequivocal evidence (symptoms or signs including performance on neuropsychological testing) of intellectual or motor impairment. The abnormal motor signs usually include slow or clumsy movements of extremities.

Stage 2 (moderate): Able to perform basic activities of self care at home but cannot work or maintain more demanding aspects of daily life (eg, maintain finances, read text more complex than a tabloid newspaper).

Stage 3 (severe): Major intellectual incapacity (cannot follow news or personal events, cannot sustain complex conversation, considerable slowing of all output) or motor disability

**CODING FOR Q45A, MYELOPATHY SEVERITY.**

**Stage 0:** Normal

**Stage 1:** Tandem gait may be impaired, but the patient can walk without assistance.

**Stage 2:** Ambulatory, but may require single prop (eg, cane).

**Stage 3:** Cannot walk unassisted, requiring walker or personal support, usually with slowing and clumsiness of arms as well.

Q47a. **NEUROLOGICAL OUTCOME**   
 1 – no need for neurological follow-up – return to regular 2-year neuropsychological testing (Form 8); 2 – neurological (Form 10) and neuropsychological (Form 8) follow-up in 6 months; 4 - other; 9 - unknown

Instructions: Record here comments about the exam. Eg, if you detect an abnormality, what do you think is its cause? Detail additional work-up here.

Q47b. **FURTHER TESTING** 1 - no; 2 - yes (select from list)

MRI	<input type="checkbox"/>	HTLV serology	<input type="checkbox"/>	Biopsy	<input type="checkbox"/>
CSF	<input type="checkbox"/>	Thyroid function	<input type="checkbox"/>	Further Neuropsych	<input type="checkbox"/>
NCV	<input type="checkbox"/>	RPR/FTA	<input type="checkbox"/>	Other	<input type="checkbox"/> _____
		B12	<input type="checkbox"/>		

Q48. **WERE YOU AWARE OF PARTICIPANT'S SEROSTATUS DURING EXAM?**   
 1 - yes; 2 - no; 9 - missing

Q49. **NEUROLOGICAL EXAMINER CODE**

Instructions: This section is for additional clinical information to be obtained after completion of the neurological history and exam. Once these sections have been completely finished and the codes recorded, move on to this section and record your notes.

Clinical symptoms: Since your last MACS visit, have you had any HIV-related illnesses, such as fever, diarrhea, PCP.

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Antiviral therapy: Describe your antiviral use: start date, type of antiviral, side effects.

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## CDC-DEFINED AIDS DIAGNOSES

- 03 **Toxoplasmosis** (at a site other than or in addition to liver, spleen, muscle or lymph nodes)
- 09 **Primary lymphoma of brain**
- 11 **Diffuse, undifferentiated B-cell non-Hodgkin's lymphoma** metastatic to brain
- 12 **Progressive multifocal leukoencephalopathy** (Papovavirus infection, brain)
- 13 **HIV encephalopathy** determined to be probable after review by Neuropsychology working group.
- 20 **Cryptococcal meningitis**
- 27 **CMV polyradiculitis.** Usually developing in a patient with advanced immune deficiency who has evidence of CMV infection elsewhere, eg, CMV retinitis, colitis, with the subacute onset of lower extremity weakness, sacral/back pain, sphincter disturbance. Cerebrospinal fluid analyses usually show a marked inflammatory response with elevated WBC, total protein, and in 50%, positive CMV culture. Autopsy confirmation may be present with demonstration of CMV in the lumbosacral nerve roots.

## MYELOPATHIES

- 3-120 **Vacuolar myelopathy.** Usually developing in a patient with advanced immune deficiency and symptomatic HIV disease, will include the development of leg weakness, usually bilateral, with increased tone, spasticity, and hyperreflexia. HIV dementia often co-exists. Imaging studies of the spinal cord (myelography, spinal MRI, spinal CT) are usually normal. Cerebrospinal fluid analysis has no specific features.
- 3-121 **Infectious causes of myelopathy.** These would include Pott's disease (tuberculosis of the spine), epidural bacterial abscesses, and herpes group infections of the spine. An example of the latter might be a patient with advanced immune deficiency with evidence of CMV elsewhere who develops acute spinal cord dysfunction and has positive spinal fluid culture for CMV.
- 1-122 **Metabolic/nutritional causes.** Example: Vitamin B12 or vitamin E deficiency.
- 1-123 **Other myelopathies,** not otherwise specified. For example, patients with cervical spondylosis, degeneration of the spine with compressive myelopathies.

## MYOPATHIES

- 3-130 **HIV-related polymyositis.** The development of weakness, principally in proximal muscle groups (thighs, shoulders) with muscle aching (myalgias), elevated levels of blood creatine phosphokinase (CPK), EMG, if performed may show evidence of a myopathy. Muscle biopsy, if performed, may show inflammatory necrosis.
- 1-131 **Toxic myopathy.** This is clinically indistinguishable from HIV-1 related polymyositis and usually occurs in patients who have received AZT at high dose (>1000 mg daily) for at least 9 to 12 months. Clinical picture and EMG cannot distinguish toxic myopathy from HIV polymyositis and we don't yet know if there are any specific biopsy findings to distinguish the two conditions. In practice, if AZT is discontinued and there is clinical improvement with reduction in the myalgias and a drop in the blood CPK levels, the myopathy is usually categorized as being a toxic effect of AZT.
- 1-132 **Other myopathies,** not otherwise specified. These might include muscular dystrophy, severe muscle wasting from nutritional deficiency.

## HIV-RELATED PERIPHERAL NEUROPATHIES

- 3-100 **Cranial neuropathies.** The acute or subacute development of cranial neuropathies thought to be related to HIV infection, eg, development of facial palsy in the setting of acute seroconversion illness with acute aseptic meningitis.
- 3-101 **Painful sensory neuropathy.** The development of painful paresthesias, dysesthesias (usually in a patient with advanced immune deficiency and symptomatic HIV disease) with objective signs of peripheral neuropathy such as depressed ankle reflexes, contact hypersensitivity in the feet, impaired vibratory sensibility in the feet. Additional supporting evidence would include nerve conduction velocities (NCV's), electromyography (EMG), and sural nerve biopsy.
- 3-102 **Inflammatory demyelinating neuropathy.** The acute or subacute development of motor weakness (usually with relatively little sensory involvement) usually in a patient in the relatively early stages of HIV infection, ie, before symptomatic HIV disease. Examination findings would include hypo- or areflexia, motor weakness, and variable sensory deficit. Corroborating evidence would include NCV's and/or EMG's demonstrating marked slowing of conduction velocities and/or denervation. Sural nerve biopsy indicating inflammation demyelination in the peripheral nerve.
- 3-103 **Mononeuritis multiplex.** Usually developing in a patient with symptomatic HIV disease with the development of multifocal signs and symptoms, eg, numbness, weakness, in the distribution of 2 or more named peripheral nerves, eg, foot drop and hand numbness.
- 3-105 **Other HIV neuropathies** (not otherwise specified). Includes all other neuropathies that might be a consequence directly or indirectly of HIV infection.

## OTHER NEUROPATHIES (NON-HIV RELATED)

- 1-110 **Cranial neuropathies.** The development of cranial neuropathies considered not to be a consequence of HIV infection. These might include development of progressive hearing loss or optic neuritis.
- 1-111 **Entrapment neuropathies.** These include the development of traumatic neuropathies affecting a named peripheral nerve with numbness, weakness, and/or pain in the distribution of the nerve, eg, carpal tunnel, tarsal tunnel, cubital tunnel.
- 1-112 **Toxic neuropathies.** These include the development of neuropathies (which are usually painful or sensory neuropathies) related to toxic effects of drugs, eg, vincristine used in the treatment of KS, or dideoxycytodine (ddC) or dideoxyinosine (ddI). Toxic neuropathies can also develop with excessive doses of vitamin B6 (pyridoxine).
- 1-113 **Diabetic neuropathy.** The development of sensory motor neuropathy, autonomic neuropathy, diabetic amyotrophy in a patient with longstanding, usually insulin-dependent, diabetes mellitus.
- 1-114 **Other neuropathies,** not otherwise specified. These might include neuropathies related to syphilis, nutritional deficiencies, alcoholism, hereditary Charcot-Marie-Tooth, meralgia paresthetica.

## OTHER NEUROLOGICAL DISEASES

- 1-140 **Neurosyphilis.** This would include a past or current history of treatment for neurosyphilis, either asymptomatic neurosyphilis (usually diagnosed if a lumbar puncture is done and the CSF VDRL is positive) or symptomatic neurosyphilis. Treatment of neurosyphilis typically includes (a) high doses of intravenous penicillin given during a 10 to 14 day hospital stay, or (b) daily doses of procaine penicillin with probenecid given for 10 to 14 days.
- 3-141 **HIV aseptic meningitis.** Development of fever, headache, neck stiffness, cranial neuropathies and mental confusion of encephalopathy. Usually associated with seroconversion illness in an otherwise well individual.
- 3-142 **Possible HIV encephalopathy.** Case reviewed by Neuropsychology Working Group (NPWG) as possible. Cases not yet reviewed by NPWG also are in this category.
- 3-144 **Herpes Zoster Meningitis.** ICD-9 code 53.0

**END OF EXAM - NEUROLOGY**

After completing the exam, the examiner should answer the following questions (these questions are for local use only – they are not sent to CAMACS):

a) Did the subject have any temporary physical limitations (such as a broken arm, swollen fingers, etc.) that might have affected the test results? (1 = yes, 2 = no) ..... \_\_\_\_\_

b) Based on the information you have available from your exam, how would you rate the overall exam? (Include information from the standard neurological exam only.)  
(1) Normal  
(2) Equivocal  
(3) Definitely abnormal  
(9) Exam not completed ..... \_\_\_\_\_

c) **Differential Diagnosis (Non-HIV):** (List all possible diagnoses *unrelated* to HIV infection. Use diagnoses on attached pages. If none, write 'NONE'.) [Coding: Enter 2 for none, otherwise leave blank] ..... \_\_\_\_\_  
  
Diff Dx (non-HIV): \_\_\_\_\_  
\_\_\_\_\_

d) **Differential Diagnosis (HIV-related):** (List all possible diagnoses related to HIV infection. Use diagnoses on attached pages. If none, write 'NONE'.) [Coding: Enter 2 for none, otherwise leave blank] ..... \_\_\_\_\_  
  
Diff Dx (HIV-related): \_\_\_\_\_  
\_\_\_\_\_

e) Relative to this subject's previous exams, the current exam shows:  
(1) global deterioration; (2) deterioration overall, though some areas were stable or showed improvement; (3) stable overall; (4) improvement overall, though some areas were stable or showed deterioration; (5) global improvement; (9) unable to evaluate ..... \_\_\_\_\_

f) Do you think this exam should be reviewed for HIV dementia? (1=Y, 2=N) ..... \_\_\_\_\_

