

INITIAL VISIT PACKET NACC UNIFORM DATA SET (UDS)

Form D1: Clinician Diagnosis

ADC name: _____ Subject ID: _____ Form date: ____ / ____ / _____
 Visit #: _____ Examiner's initials: _____

INSTRUCTIONS: This form is to be completed by the clinician. For additional clarification and examples, see UDS Coding Guidebook for Initial Visit Packet, Form D1. Check only one box per question.

This form is divided into three main sections:

- Section 1 **Cognitive and behavioral status:** Normal cognition / MCI / dementia and dementia syndrome
- Section 2 **Biomarkers, imaging, and genetics:** Neurodegenerative imaging and CSF biomarkers, imaging evidence for CVD, and known genetic mutations for AD and FTLD
- Section 3 **Etiological diagnoses:** presumed etiological diagnoses for the cognitive disorder

1. Diagnosis method — responses in this form are based on diagnosis by:

- 1 A single clinician 2 A formal consensus panel 3 Other (e.g., two or more clinicians or other informal group)

SECTION 1: Cognitive and behavioral status

2. Does the subject have normal cognition (global CDR=0 and/or neuropsychological testing within normal range) and normal behavior (i.e., the subject does not exhibit behavior sufficient to diagnose MCI or dementia due to FTLD or LBD)?

- 0 No (**CONTINUE TO QUESTION 3**)
 1 Yes (**SKIP TO QUESTION 6**)

ALL-CAUSE DEMENTIA

The subject has cognitive or behavioral (neuropsychiatric) symptoms that meet all of the following criteria:

- Interfere with ability to function as before at work or at usual activities?
- Represent a decline from previous levels of functioning?
- Are not explained by delirium or major psychiatric disorder?
- Include cognitive impairment detected and diagnosed through a combination of 1) history-taking and 2) objective cognitive assessment (bedside or neuropsychological testing)?

AND

Impairment in one* or more of the following domains.

- Impaired ability to acquire and remember new information
- Impaired reasoning and handling of complex tasks, poor judgment
- Impaired visuospatial abilities
- Impaired language functions
- Changes in personality, behavior, or comporment

** In the event of single-domain impairment (e.g., language in PPA, behavior in bvFTD, posterior cortical atrophy), the subject must not fulfill criteria for MCI.*

3. Does the subject meet the criteria for dementia?

- 0 No (**SKIP TO QUESTION 5**)
 1 Yes (**CONTINUE TO QUESTION 4**)

4. If the subject meets criteria for dementia, answer Questions 4a–4f below and then SKIP TO QUESTION 6.

Based entirely on the history and examination (including neuropsychological testing), what is the cognitive/behavioral syndrome? **Select one or more as Present; all others will default to Absent in the NACC database.**

| Dementia syndrome | Present |
|--|----------------------------|
| 4a. Amnestic multidomain dementia syndrome | <input type="checkbox"/> 1 |
| 4b. Posterior cortical atrophy syndrome (or primary visual presentation) | <input type="checkbox"/> 1 |
| 4c. Primary progressive aphasia (PPA) syndrome | <input type="checkbox"/> 1 |
| 4c1. <input type="checkbox"/> 1 Meets criteria for semantic PPA <input type="checkbox"/> 2 Meets criteria for logopenic PPA <input type="checkbox"/> 3 Meets criteria for nonfluent/agrammatic PPA <input type="checkbox"/> 4 PPA other/not otherwise specified | |
| 4d. Behavioral variant FTD (bvFTD) syndrome | <input type="checkbox"/> 1 |
| 4e. Lewy body dementia syndrome | <input type="checkbox"/> 1 |
| 4f. Non-amnestic multidomain dementia, not PCA, PPA, bvFTD, or DLB syndrome | <input type="checkbox"/> 1 |

5. If the subject does not have normal cognition or behavior and is not clinically demented, indicate the type of cognitive impairment below.

MCI CORE CLINICAL CRITERIA

- Is the subject, the co-participant, or a clinician concerned about a change in cognition compared to the subject's previous level?
- Is there impairment in one or more cognitive domains (memory, language, executive function, attention, and visuospatial skills)?
- Is there largely preserved independence in functional abilities (no change from prior manner of functioning or uses minimal aids or assistance)?

Select one syndrome from 5a–5e as being Present (all others will default to Absent in the NACC database), and then **CONTINUE TO QUESTION 6**. If you select MCI below, it should meet the MCI core clinical criteria outlined above.

| Type | Present | Affected domains | No | Yes |
|--|----------------------------|---|--|--|
| 5a. Amnestic MCI, single domain (aMCI SD) | <input type="checkbox"/> 1 | | | |
| 5b. Amnestic MCI, multiple domains (aMCI MD) | <input type="checkbox"/> 1 | CHECK YES for at least one additional domain (besides memory): 5b1. Language 5b2. Attention 5b3. Executive 5b4. Visuospatial | <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 | <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 |

Select one syndrome from 5a–5e as being Present (all others will default to Absent in the NACC database), and then **CONTINUE TO QUESTION 6**. If you select MCI below, it should meet the MCI core clinical criteria outlined above.

| Type | Present | Affected domains | No | Yes |
|--|----------------------------|---|--|--|
| 5c. Non-amnesic MCI, single domain (naMCI SD) | <input type="checkbox"/> 1 | CHECK YES to indicate the affected domain: 5c1. Language 5c2. Attention 5c3. Executive 5c4. Visuospatial | <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 | <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 |
| 5d. Non-amnesic MCI, multiple domains (naMCI MD) | <input type="checkbox"/> 1 | CHECK YES for at least two domains: 5d1. Language 5d2. Attention 5d3. Executive 5d4. Visuospatial | <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 <input type="checkbox"/> 0 | <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 <input type="checkbox"/> 1 |
| 5e. Cognitively impaired, not MCI | <input type="checkbox"/> 1 | | | |

SECTION 2: Biomarkers, imaging, and genetics

Section 2 must be completed for all subjects.

6. Indicate neurodegenerative biomarker status, using local standards for positivity.

| Biomarker findings | No | Yes | Unknown/ not assessed |
|---|----------------------------|----------------------------|----------------------------|
| 6a. Abnormally elevated amyloid on PET | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6b. Abnormally low amyloid in CSF | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6c. FDG-PET pattern of AD | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6d. Hippocampal atrophy | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6e. Tau PET evidence for AD | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6f. Abnormally elevated CSF tau or ptau | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6g. FDG-PET evidence for frontal or anterior temporal hypometabolism for FTLD | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6h. Tau PET evidence for FTLD | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6i. Structural MR evidence for frontal or anterior temporal atrophy for FTLD | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6j. Dopamine transporter scan (DATscan) evidence for Lewy body disease | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 6k. Other (SPECIFY): _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |

7. Is there evidence for cerebrovascular disease (CVD) on imaging?

| Imaging findings | No | Yes | Unknown/ not assessed |
|--|----------------------------|----------------------------|----------------------------|
| 7a. Large vessel infarct(s) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 7b. Lacunar infarct(s) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 7c. Macrohemorrhage(s) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 7d. Microhemorrhage(s) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 7e. Moderate white-matter hyperintensity (CHS score 5–6) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |
| 7f. Extensive white-matter hyperintensity (CHS score 7–8+) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 8 |

8. Does the subject have a dominantly inherited AD mutation (PSEN1, PSEN2, APP)?

0 No 1 Yes 9 Unknown/not assessed

9. Does the subject have a hereditary FTLN mutation (e.g., GRN, VCP, TARBP, FUS, C9orf72, CHMP2B, MAPT)?

0 No 1 Yes 9 Unknown/not assessed

10. Does the subject have a hereditary mutation other than an AD or FTLN mutation?

0 No 1 Yes (SPECIFY): _____ 9 Unknown/not assessed

SECTION 3: Etiologic diagnoses

Section 3 must be filled out for all subjects. Indicate presumptive etiologic diagnoses of the cognitive disorder and whether a given diagnosis is a primary, contributing, or non-contributing cause of the observed impairment, based on the clinician’s best judgment. **Select one or more diagnoses as Present; all others will default to Absent in the NACC database.** Only one diagnosis should be selected as 1=Primary.

For subjects with normal cognition: Indicate the presence of any diagnoses by marking Present, and leave the questions on whether the diagnosis was primary, contributing, or non-contributing blank. Subjects with positive biomarkers but no clinical symptoms of Alzheimer’s disease, Lewy body disease, or frontotemporal lobar degeneration **should not** have these diagnoses marked as Present. Instead, the biomarker data from Section 2 can be used to identify the presence of preclinical disease.

| Etiologic diagnoses | Present | Primary | Contributing | Non-contributing |
|--|----------------------------|---------------------------------|----------------------------|----------------------------|
| 11. Alzheimer’s disease | <input type="checkbox"/> 1 | 11a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 12. Lewy body disease 12b. <input type="checkbox"/> 1 Parkinson’s disease | <input type="checkbox"/> 1 | 12a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 13. Multiple system atrophy | <input type="checkbox"/> 1 | 13a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 14. Frontotemporal lobar degeneration | | | | |
| 14a. Progressive supranuclear palsy (PSP) | <input type="checkbox"/> 1 | 14a1 <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 14b. Corticobasal degeneration (CBD) | <input type="checkbox"/> 1 | 14b1 <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 14c. FTLN with motor neuron disease | <input type="checkbox"/> 1 | 14c1 <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 14d. FTLN NOS | <input type="checkbox"/> 1 | 14d1 <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 14e. If FTLN (Questions 14a – 14d) is Present, specify FTLN subtype: <input type="checkbox"/> 1 Tauopathy <input type="checkbox"/> 2 TDP-43 proteinopathy <input type="checkbox"/> 3 Other (SPECIFY): _____ <input type="checkbox"/> 9 Unknown | | | | |

SECTION 3: Etiologic diagnoses (cont.)

Section 3 must be filled out for all subjects. Indicate presumptive etiologic diagnoses of the cognitive disorder and whether a given diagnosis is a primary, contributing, or non-contributing cause of the observed impairment, based on the clinician's best judgment. **Select one or more diagnoses as Present; all others will default to Absent in the NACC database.** Only one diagnosis should be selected as **1=Primary**.

For subjects with normal cognition: Indicate the presence of any diagnoses by selecting **1=Present**, and leave the questions on whether the diagnosis was primary, contributing, or non-contributing blank. Subjects with positive biomarkers but no clinical symptoms of Alzheimer's disease, Lewy body disease, or frontotemporal lobar degeneration **should not** have these diagnoses selected as Present. Instead, the biomarker data from Section 2 can be used to identify the presence of preclinical disease.

| Etiologic diagnoses | | Present | Primary | Contributing | Non-contributing |
|---------------------|---|----------------------------|--------------------------------|----------------------------|----------------------------|
| 15. | Vascular brain injury (based on clinical or imaging evidence) <i>If significant vascular brain injury is absent, SKIP TO QUESTION 16.</i> | <input type="checkbox"/> 1 | 15a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 15b. | Previous symptomatic stroke? <input type="checkbox"/> 0 No (SKIP TO QUESTION 15c) <input type="checkbox"/> 1 Yes | | | | |
| 15b1. | Temporal relationship between stroke and cognitive decline? <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes | | | | |
| 15b2. | Confirmation of stroke by neuroimaging? <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown; no relevant imaging data available | | | | |
| 15c. | Is there imaging evidence of cystic infarction in cognitive network(s)? <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown; no relevant imaging data available | | | | |
| 15d. | Is there imaging evidence of cystic infarction, imaging evidence of extensive white matter hyperintensity (CHS grade 7–8+), <u>and</u> impairment in executive function? <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown; no relevant imaging data available | | | | |
| 16. | Essential tremor | <input type="checkbox"/> 1 | 16a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 17. | Down syndrome | <input type="checkbox"/> 1 | 17a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 18. | Huntington's disease | <input type="checkbox"/> 1 | 18a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 19. | Prion disease (CJD, other) | <input type="checkbox"/> 1 | 19a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |

| Etiologic diagnoses | | Present | Primary | Contributing | Non-contributing |
|---------------------|--|----------------------------|--------------------------------|----------------------------|----------------------------|
| 20. | Traumatic brain injury 20b. If Present, does the subject have symptoms consistent with chronic traumatic encephalopathy? <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown | <input type="checkbox"/> 1 | 20a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 21. | Normal-pressure hydrocephalus | <input type="checkbox"/> 1 | 21a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 22. | Epilepsy | <input type="checkbox"/> 1 | 22a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 23. | CNS neoplasm 23b. <input type="checkbox"/> 1 Benign <input type="checkbox"/> 2 Malignant | <input type="checkbox"/> 1 | 23a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 24. | Human immunodeficiency virus (HIV) | <input type="checkbox"/> 1 | 24a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 25. | Cognitive impairment due to other neurologic, genetic, or infectious conditions not listed above 25b. If Present, specify: _____ | <input type="checkbox"/> 1 | 25a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |

Section 3 must be filled out for all subjects. Indicate presumptive etiologic diagnoses of the cognitive disorder and whether a given diagnosis is a primary, contributing, or non-contributing cause of the observed impairment, based on the clinician's best judgment. **Select one or more diagnoses as Present; all others will default to Absent in the NACC database.** Only one diagnosis should be selected as **1= Primary**.

For subjects with normal cognition: Indicate the presence of any diagnoses by selecting **1=Present**, and leave the questions on whether the diagnosis was primary, contributing, or non-contributing blank. Subjects with positive biomarkers but no clinical symptoms of Alzheimer's disease, Lewy body disease, or frontotemporal lobar degeneration **should not** have these diagnoses selected as Present. Instead, the biomarker data from Section 2 can be used to identify the presence of preclinical disease.

| Condition | Present | Primary | Contributing | Non-contributing |
|---|----------------------------|--------------------------------|----------------------------|----------------------------|
| 26. Active depression 26b. If Present, select one: <input type="checkbox"/> 0 Untreated <input type="checkbox"/> 1 Treated with medication and/or counseling | <input type="checkbox"/> 1 | 26a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 27. Bipolar disorder | <input type="checkbox"/> 1 | 27a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 28. Schizophrenia or other psychosis | <input type="checkbox"/> 1 | 28a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 29. Anxiety disorder | <input type="checkbox"/> 1 | 29a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 30. Delirium | <input type="checkbox"/> 1 | 30a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 31. Post-traumatic stress disorder (PTSD) | <input type="checkbox"/> 1 | 31a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 32. Other psychiatric disease 32b. If Present, specify: _____ | <input type="checkbox"/> 1 | 32a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |

| | | | | |
|---|----------------------------|--------------------------------|----------------------------|----------------------------|
| 33. Cognitive impairment due to alcohol abuse 33b. Current alcohol abuse: <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown | <input type="checkbox"/> 1 | 33a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 34. Cognitive impairment due to other substance abuse | <input type="checkbox"/> 1 | 34a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 35. Cognitive impairment due to systemic disease/ medical illness (as indicated on Form D2) | <input type="checkbox"/> 1 | 35a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 36. Cognitive impairment due to medications | <input type="checkbox"/> 1 | 36a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 37. Cognitive impairment NOS 37b. If Present, specify: _____ | <input type="checkbox"/> 1 | 37a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 38. Cognitive impairment NOS 38b. If Present, specify: _____ | <input type="checkbox"/> 1 | 38a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |
| 39. Cognitive impairment NOS 39b. If Present, specify: _____ | <input type="checkbox"/> 1 | 39a <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 |