TELEPHONE INITIAL VISIT PACKET NACC UNIFORM DATA SET (UDS)

Form D1: Clinician Diagnosis

CLEAR FORM

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 Telephone 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 comportment

** 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	☐ 1
4b. Posterior cortical atrophy syndrome (or primary visual presentation)	☐ 1
4c. Primary progressive aphasia (PPA) syndrome 4c1. ☐ 1 Meets criteria for semantic PPA ☐ 2 Meets criteria for logopenic PPA ☐ 3 Meets criteria for nonfluent/agrammatic PPA ☐ 4 PPA other/not otherwise specified	☐ 1
4d. Behavioral variant FTD (bvFTD) syndrome	☐ 1
4e. Lewy body dementia syndrome	☐ 1
4f. Non-amnestic multidomain dementia, not PCA, PPA, bvFTD, or DLB syndrome	☐ 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)	☐ 1			
5b. Amnestic MCI, multiple domains (aMCI MD)	☐ 1	**CHECK YES for at least one additional domain (besides memory):** 5b1. Language 5b2. Attention 5b3. Executive 5b4. Visuospatial	 ☐ 0 ☐ 0 ☐ 0 ☐ 0	 ☐ 1 ☐ 1 ☐ 1 ☐ 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-amnestic MCI, single domain (naMCI SD)	☐ 1	**CHECK YES to indicate the affected domain:** 5c1. Language 5c2. Attention 5c3. Executive 5c4. Visuospatial	 ☐ 0 ☐ 0 ☐ 0 ☐ 0	 ☐ 1 ☐ 1 ☐ 1 ☐ 1
5d. Non-amnestic MCI, multiple domains (naMCI MD)	☐ 1	**CHECK YES for at least two domains:** 5d1. Language 5d2. Attention 5d3. Executive 5d4. Visuospatial	 ☐ 0 ☐ 0 ☐ 0 ☐ 0	 ☐ 1 ☐ 1 ☐ 1 ☐ 1
5e. Cognitively impaired, not MCI	☐ 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	☐ 0	☐ 1	☐ 8
6b. Abnormally low amyloid in CSF	☐ 0	☐ 1	☐ 8
6c. FDG-PET pattern of AD	☐ 0	☐ 1	☐ 8
6d. Hippocampal atrophy	☐ 0	☐ 1	☐ 8
6e. Tau PET evidence for AD	☐ 0	☐ 1	☐ 8
6f. Abnormally elevated CSF tau or ptau	☐ 0	☐ 1	☐ 8
6g. FDG-PET evidence for frontal or anterior temporal hypometabolism for FTLD	☐ 0	☐ 1	☐ 8
6h. Tau PET evidence for FTLD	☐ 0	☐ 1	☐ 8
6i. Structural MR evidence for frontal or anterior temporal atrophy for FTLD	☐ 0	☐ 1	☐ 8
6j. Dopamine transporter scan (DATscan) evidence for Lewy body disease	☐ 0	☐ 1	☐ 8
6k. Other (SPECIFY): _____	☐ 0	☐ 1	

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

Imaging findings	No	Yes	Unknown/ not assessed
7a. Large vessel infarct(s)	☐ 0	☐ 1	☐ 8
7b. Lacunar infarct(s)	☐ 0	☐ 1	☐ 8
7c. Macrohemorrhage(s)	☐ 0	☐ 1	☐ 8
7d. Microhemorrhage(s)	☐ 0	☐ 1	☐ 8
7e. Moderate white-matter hyperintensity (CHS score 5–6)	☐ 0	☐ 1	☐ 8
7f. Extensive white-matter hyperintensity (CHS score 7–8+)	☐ 0	☐ 1	☐ 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 FTLD 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 FTLD 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	☐ 1	11a ☐ 1	☐ 2	☐ 3
12. Lewy body disease 12b. ☐ 1 Parkinson's disease	☐ 1	12a ☐ 1	☐ 2	☐ 3
13. Multiple system atrophy	☐ 1	13a ☐ 1	☐ 2	☐ 3
14. Frontotemporal lobar degeneration				
14a. Progressive supranuclear palsy (PSP)	☐ 1	14a1 ☐ 1	☐ 2	☐ 3
14b. Corticobasal degeneration (CBD)	☐ 1	14b1 ☐ 1	☐ 2	☐ 3
14c. FTLD with motor neuron disease	☐ 1	14c1 ☐ 1	☐ 2	☐ 3
14d. FTLD NOS	☐ 1	14d1 ☐ 1	☐ 2	☐ 3
14e. If FTLD (Questions 14a – 14d) is Present, specify FTLD subtype: ☐ 1 Tauopathy ☐ 2 TDP-43 proteinopathy ☐ 3 Other (SPECIFY): _____ ☐ 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) *If significant vascular brain injury is absent,* **SKIP TO QUESTION 16.** 15b. Previous symptomatic stroke? ☐ 0 No **(SKIP TO QUESTION 15c)** ☐ 1 Yes 15b1. Temporal relationship between stroke and cognitive decline? ☐ 0 No ☐ 1 Yes 15b2. Confirmation of stroke by neuroimaging? ☐ 0 No ☐ 1 Yes ☐ 9 Unknown; no relevant imaging data available 15c. Is there imaging evidence of cystic infarction in cognitive network(s)? ☐ 0 No ☐ 1 Yes ☐ 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+), and impairment in executive function? ☐ 0 No ☐ 1 Yes ☐ 9 Unknown; no relevant imaging data available	☐ 1	15a ☐ 1	☐ 2	☐ 3
16. Essential tremor	☐ 1	16a ☐ 1	☐ 2	☐ 3
17. Down syndrome	☐ 1	17a ☐ 1	☐ 2	☐ 3
18. Huntington's disease	☐ 1	18a ☐ 1	☐ 2	☐ 3
19. Prion disease (CJD, other)	☐ 1	19a ☐ 1	☐ 2	☐ 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? ☐ 0 No ☐ 1 Yes ☐ 9 Unknown	☐ 1	20a ☐ 1	☐ 2	☐ 3
21.	Normal-pressure hydrocephalus	☐ 1	21a ☐ 1	☐ 2	☐ 3
22.	Epilepsy	☐ 1	22a ☐ 1	☐ 2	☐ 3
23.	CNS neoplasm 23b. ☐ 1 Benign ☐ 2 Malignant	☐ 1	23a ☐ 1	☐ 2	☐ 3
24.	Human immunodeficiency virus (HIV)	☐ 1	24a ☐ 1	☐ 2	☐ 3
25.	Cognitive impairment due to other neurologic, genetic, or infectious conditions not listed above 25b. If Present, specify: _____	☐ 1	25a ☐ 1	☐ 2	☐ 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: ☐ 0 Untreated ☐ 1 Treated with medication and/or counseling	☐ 1	26a ☐ 1	☐ 2	☐ 3
27.	Bipolar disorder	☐ 1	27a ☐ 1	☐ 2	☐ 3
28.	Schizophrenia or other psychosis	☐ 1	28a ☐ 1	☐ 2	☐ 3
29.	Anxiety disorder	☐ 1	29a ☐ 1	☐ 2	☐ 3
30.	Delirium	☐ 1	30a ☐ 1	☐ 2	☐ 3
31.	Post-traumatic stress disorder (PTSD)	☐ 1	31a ☐ 1	☐ 2	☐ 3
32.	Other psychiatric disease 32b. If Present, specify: _____	☐ 1	32a ☐ 1	☐ 2	☐ 3

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