UDS3 Diagnostic Issues

Form D1's form

Caveat: assumes that **cognition** is the principal diagnostic axis

- Section 1 Cognitive/Behavioral Syndrome
  - Normal
  - MCI
  - Dementia
- Section 2 Biomarkers
  - Imaging & Fluid
  - Genetics
- Etiological diagnoses

D1. Diagnosis: Syndromic Dementia

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

Form D1: Biomarkers

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	

Breakdown of Syndrome x Etiology

	Clinically Defined Syndromes				
PATHOLOGIC Diagnoses ↓	Amnestic Multi Domain Dementia	PPA syndrome	bvFTD	LBD syndrome	Non-AMN DEM not PCA, PPA, etc
Alzheimer	**1015**	**33**	3	2	6
Lewy Body	5	0	0	**42**	1
PSP/CBD/FTLD w/ MND	9	**6**	**8**	0	16
FTLD No MND	5	**81**	**83**	0	4
Vascular	25	0	0	0	2
Other	26	0	1	0	4

Low rate of ascertainment of biomarkers

Final Etiological Diagnoses	Total	Biomarkers Assessed*
Normal Cognition	3086	180
AD spectrum (MCI, DEM)	1649	198
LBD spectrum (MCI, DEM)	89	17
All FTLD spectrum (MCI, DEM)	312	81
Cerebrovascular	103	14

*Assessed means that one of Amyloid PET or Amyloid CSF or Tau PET or Tau CSF was assessed

Cerebrovascular Imaging Biomarkers

	Total	Normal	Demented	MCI
7a. Large vessel infarcts				
No	1564	515	691	306
Yes	39	3	21	11
Unknown/not assessed	4094	2568	798	525
7b. Lacunar infarcts				
No	1484	493	664	282
Yes	119	26	52	34
Unknown/not assessed	4094	2567	794	526
7c. Macrohemorrhages				
No	1571	511	698	311
Yes	6	2	2	1
Unknown/not assessed	4120	2573	810	530
7d. Microhemorrhages				
No	1251	348	597	261
Yes	43	8	21	12
Unknown/not assessed	4403	2730	892	569
7e. Moderate white matter hyperintensity				
No	1402	474	614	267
Yes	170	29	89	48
Unknown/not assessed	4125	2583	807	527
7f. Extensive white matter hyperintensity				
No	1527	499	675	303
Yes	47	11	24	11
Unknown/not assessed	4123	2576	811	528

D1. Diagnosis: Neurodegenerative Etiologies

SECTION 3: Etiologic diagnoses

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

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.**	☐ 1	15a ☐ 1	☐ 2	☐ 3
15b. Previous symptomatic stroke? ☐ 0 No **(SKIP TO QUESTION 15c)** ☐ 1 Yes	Collected in UDS2 B2 (HIS and CVD)			
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	Collected in UDS2 B2 (HIS and CVD)			
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				

D1. Diagnosis: Etiology