NACC UNIFORM DATA SET **LBD MODULE**

Coding Guidebook for IVP

LBD Module, August 2017
UDS Version 3.0, March 2015

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Revisions made to this Guidebook since LBD Module implementation (August 2017)

Date yyyy-mm-dd	Description	Form(s) affected	Question(s) affected	Data element(s) affected
2018-06-04	Clarification added for B9L, questions 2 – 5	B9L	2 – 5	N/A
2017-10-23	Item descriptions on form clarified and made consistent with Noise Pareidolia worksheet	C1L	2a – 2d	LBNPFACE, LBNPNOIS, LBNPTCOR, LBNPPARD
2017-09-19	LBD diagnostic criteria updated to reflect 2017 guidelines of Dementia With Lewy Bodies Consortium	D1L	6	LBCOGDX

Introduction

The LBD Module to the Uniform Data Set (UDS) is designed for:

- Persons with a diagnosis of dementia with Lewy bodies (DLB) or Parkinson's disease dementia (PDD)
- Persons with mild cognitive impairment who are thought to be at risk for developing DLB or PD
- Control and Alzheimer's disease participants, at the Center's discretion

Generally, it will be unnecessary to complete an LBD Module and FTLD Module for a given participant.

IMPORTANT NOTES

- **Timing** — The LBD Module evaluation is intended to be completed as part of a UDS visit. If the UDS evaluation and the LBD evaluation are separated into two days, please complete the LBD evaluation within two weeks of the UDS evaluation.
- **Visit number** — Even when the visit is split into two days, the same visit number MUST be used in the form header on all forms in both packets (UDS and LBD) from both days.
- **IVP vs FVP** — When a UDS enrollee is being given the LBD evaluation for the first time, you should use the LBD Module Initial Visit Packet, even if you are using the UDS Follow-up Visit Packet.
- **Data inconsistencies within the LBD Module**: Any data inconsistencies between forms of the LBD Module must be corrected before the packet will be accepted into the NACC Database. For example, if the LBD Module indicates the presence of a severe gait disorder on Form B3L — UPDRS Part III, gait abnormality should not be indicated as absent on Form D1L — Clinical DLB and PD Features.
- **Data inconsistencies between the LBD Module and the UDS** — Any data inconsistencies between the LBD Module and the UDS must be corrected before the packets will be accepted into the NACC database. For example, if the LBD Module indicates the presence of REM sleep behavior disorder, it should not be indicated as absent in the UDS.

Form B1L: Clinical Symptoms and Exam

AUTONOMIC SYMPTOMS CHECKLIST

In the past six months …	No	Yes	Unknown
Questions 1 through 19, below: Make every effort to answer 1=Yes or 0=No using the information provided by the participant and co-participant. Indicate 9=Unknown when it is not possible to obtain information regarding the particular symptom from the participant and co-participant.			
1. Does the participant dribble saliva during the day?	☐ 0	☐ 1	☐ 9
2. Does the participant have difficulty swallowing?	☐ 0	☐ 1	☐ 9
3. Does the participant have altered interest in sex?	☐ 0	☐ 1	☐ 9
4. Does the participant have problems having sex?	☐ 0	☐ 1	☐ 9
5. Does the participant have a recent change in weight (not related to dieting)?	☐ 0	☐ 1	☐ 9
6. Does the participant report a change in the ability to taste or smell?	☐ 0	☐ 1	☐ 9
7. Does the participant experience excessive sweating (not related to hot weather)?	☐ 0	☐ 1	☐ 9
8. Does the participant report having difficulty tolerating cold weather?	☐ 0	☐ 1	☐ 9
9. Does the participant report having difficulty tolerating hot weather?	☐ 0	☐ 1	☐ 9
10. Does the participant experience double vision (two separate real objects, and not blurred vision)?	☐ 0	☐ 1	☐ 9
11. Does the participant have problems with constipation?	☐ 0	☐ 1	☐ 9
12. Does the participant have to strain to pass hard stools?	☐ 0	☐ 1	☐ 9
13. Has the participant had involuntary loss of stools?	☐ 0	☐ 1	☐ 9
14. Has the participant had the feeling that after passing urine, their bladder was not completely empty?	☐ 0	☐ 1	☐ 9
15. Has the participant's stream of urine been weak or reduced?	☐ 0	☐ 1	☐ 9
16. Has the participant had to pass urine within two hours of the previous urination?	☐ 0	☐ 1	☐ 9
17. Has the participant complained of feeling light-headed or dizzy when standing up?	☐ 0	☐ 1	☐ 9
18. Has the participant become light-headed after standing for some time?	☐ 0	☐ 1	☐ 9
19. Has the participant fainted?	☐ 0	☐ 1	☐ 9

20. Indicate the first predominant symptom to appear during the participant's lifetime: (SELECT ONLY ONE)

- ☐ 1 Dribbling saliva during the day
- ☐ 2 Difficulty swallowing
- ☐ 3 Altered interest in sex
- ☐ 4 Problems having sex
- ☐ 5 Recent change in weight not related to dieting
- ☐ 6 Change in the ability to taste or smell
- ☐ 7 Excessive sweating
- ☐ 8 Difficulty tolerating cold weather
- ☐ 9 Difficulty tolerating hot weather
- ☐ 10 Double vision
- ☐ 11 Constipation
- ☐ 12 Straining to pass hard stools
- ☐ 13 Involuntary loss of stools
- ☐ 14 Feeling after passing urine that bladder is not completely empty
- ☐ 15 Stream of urine weak or reduced
- ☐ 16 Passing urine within two hours of previous urination
- ☐ 17 Feeling light-headed or dizzy when standing up
- ☐ 18 Feeling light-headed after standing for some time
- ☐ 19 Fainting
- ☐ 88 Not applicable — never experienced any of these symptoms
- ☐ 99 Unknown

Question 20: Indicate the first predominant autonomic symptom, which could have occurred a number of years before this visit (i.e., irrespective of whether the participant has experienced any autonomic symptoms in the past six months).

21. At what age did the first predominant symptom appear? ___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 21: This question refers to the age of onset of the first predominant autonomic symptom, which could have occurred a number of years prior to this visit (i.e., irrespective of whether the participant has experienced any autonomic symptoms in the past six months).

If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of the first predominant autonomic symptom. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced the autonomic symptoms listed in Questions 1 – 19.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

MEASUREMENTS		
Supine position	22. Systolic blood pressure:	___ ___ ___ *(888=Not assessed)*
	23. Diastolic blood pressure:	___ ___ ___ *(888=Not assessed)*
	24. Heart rate:	___ ___ ___ *(888=Not assessed)*
Standing position	25. Systolic blood pressure:	___ ___ ___ *(888=Not assessed)*
	26. Diastolic blood pressure:	___ ___ ___ *(888=Not assessed)*
	27. Heart rate:	___ ___ ___ *(888=Not assessed)*

Questions 22, 23, 25, and 26: Wait at least 1 minute before taking the indicated blood pressure measurement.

Questions 24 and 27: Wait at least 1 minute before taking the heart rate measurement.

AGE OF ONSET OF NON-MOTOR SYMPTOMS

28. Age of onset of probable REM sleep behavior disorder:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 28: If the participant has core features consistent with a diagnosis of probable RBD (i.e., dream enactment behavior with or without polysomnography confirmation), provide the age of onset. If the participant has never exhibited features consistent with probable RBD, indicate 888=Not applicable.

The clinician must use his/her best judgment to estimate the age of onset of probable RBD if the exact age is unknown. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

29. Age of onset of impaired smell:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 29: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of impaired smell. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced impaired smell.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

AGE OF ONSET OF MOTOR SYMPTOMS

30. Age of onset of gait disorder:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 30: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of gait disorder. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced a gait disorder.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

31. Age of onset of falls:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 31: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of falls. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced falls.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

32. Age of onset of tremor:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 32: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of tremor. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced tremor.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

33. Age of onset of bradykinesia:	___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 33: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of bradykinesia. For example, if the co-participant indicates that onset began in the participant's 50s, an estimated age of 55 years could be provided.

Indicate 888=Not applicable if the subject has never experienced bradykinesia.

Only indicate 999=Unknown if the medical records and participant and co-participant report are not adequate for estimating an age of onset.

34. WAS A STANDARDIZED SCALE OF AUTONOMIC SYMPTOMS COMPLETED AT THIS VISIT?

☐ 0 No **(END FORM HERE)**

☐ 1 Yes **(CONTINUE TO QUESTIONS 34a and 34b)**

34a. If yes, which version?

☐ 1 NMSS

☐ 2 SCOPA-AUT

☐ 8 Other (SPECIFY): _____

34b. If yes, what was the score? _____ *(999 = Unknown)*

If a standardized scale of autonomic symptoms was completed but the score is not available or unknown, indicate 999=Unknown.

Form B2L: UPDRS Part II — Activities of Daily Living (Determine for "On/Off")[1]

This Form uses a 5-point scale to record historical information on how the participant functions in various activities. **Speech**, **tremor**, and **walking** are items in this section and again in Form B3L, which records the evaluation of **speech**, **tremor**, and **gait** as seen by the examiner. The items **salivation** and **handwriting** also are scored as interpreted by the participant, and are not the result of the motor examination by the examiner. This differs from previous scales. Placing **handwriting** as part of history rather than direct examination was for expediency, saving the examiner time. Many examiners may prefer to obtain a sample of handwriting for their scrutiny and placement in the participant's chart as a good method of following participants.

The item **falling** reflects falling due to gait, posture, and postural reflex impairment, and not due to the "freezing" phenomenon or postural hypotension. There is a separate item to quantitate **freezing when walking**, which in its severest form causes falling, thereby receiving the maximum score. If the participant does not walk at all, please indicate 8=Not applicable for **freezing when walking**.

Since some participants with PD have sensory symptoms, the developers incorporated the item **sensory complaints related to parkinsonism** into this section. This item, as well as freezing and falling, is a new feature and is not seen in previously published rating scales. It is important to point out that there are many causes of sensory complaints, including arthritis, diabetes, and compressed spinal roots, and it is therefore important to score this item only when the examiner is certain that there are no other causes of sensory symptoms other than PD.

The historical ADL data should be obtained for participants at their best and at their worst, i.e., when "on" and when "off," for those who have clinical fluctuations from levodopa therapy.

If the participant was not able to perform a given task before onset of parkinsonian symptoms, and thus the question cannot be answered, indicate 8=Not applicable. If it is not possible to answer the question due to insufficient information from the participant and co-participant, indicate 9=Unknown.

1. Speech

- ☐ 0 Normal.
- ☐ 1 Mildly affected. No difficulty being understood.
- ☐ 2 Moderately affected. Sometimes asked to repeat statements.
- ☐ 3 Severely affected. Frequently asked to repeat statements.
- ☐ 4 Unintelligible most of the time.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

[1]Fahn S, Elton RL, UPDRS Development Committee. The Unified Parkinson's Disease Rating Scale. In Fahn S, Marsden CD, Calne DB, Goldstein M, eds. Recent developments in Parkinson's disease, Vol. 2. Florham Park, NJ: Macmillan Healthcare Information, 1987:153-163, 293-304. Reproduced by permission of the author.

2. Salivation

- ☐ 0 Normal.
- ☐ 1 Slight but definite excess of saliva in mouth; may have night time drooling.
- ☐ 2 Moderately excessive saliva; may have minimal drooling.
- ☐ 3 Marked excess of saliva with some drooling.
- ☐ 4 Marked drooling, requires constant tissue or handkerchief.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

3. Swallowing

- ☐ 0 Normal.
- ☐ 1 Rare choking.
- ☐ 2 Occasional choking.
- ☐ 3 Requires soft food.
- ☐ 4 Requires NG tube or gastrotomy feeding.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

4. Handwriting

- ☐ 0 Normal.
- ☐ 1 Slightly slow or small.
- ☐ 2 Moderately slow or small; all words are legible.
- ☐ 3 Severely affected; not all words are legible.
- ☐ 4 The majority of words are not legible.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

5. Cutting food and handling utensils

- ☐ 0 Normal.
- ☐ 1 Somewhat slow and clumsy, but no help needed.
- ☐ 2 Can cut most foods, although clumsy and slow; some help needed.
- ☐ 3 Food must be cut by someone, but can still feed slowly.
- ☐ 4 Needs to be fed.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

6. Dressing

- ☐ 0 Normal.
- ☐ 1 Somewhat slow, but no help needed.
- ☐ 2 Occasional assistance with buttoning, getting arms in sleeves.
- ☐ 3 Considerable help required, but can do some things alone.
- ☐ 4 Helpless.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

7. Hygiene

- ☐ 0 Normal.
- ☐ 1 Somewhat slow, but no help needed.
- ☐ 2 Needs help to shower or bathe; or very slow in hygienic care.
- ☐ 3 Requires assistance for washing, brushing teeth, combing hair, going to bathroom.
- ☐ 4 Foley catheter or other mechanical aids.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

8. Turning in bed and adjusting bedclothes

- ☐ 0 Normal.
- ☐ 1 Somewhat slow and clumsy, but no help needed.
- ☐ 2 Can turn alone or adjust sheets, but with great difficulty.
- ☐ 3 Can initiate, but not turn or adjust sheets alone.
- ☐ 4 Helpless.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

9. Falling (unrelated to freezing)

- ☐ 0 None.
- ☐ 1 Rare falling.
- ☐ 2 Occasionally falls, less than once per day.
- ☐ 3 Falls an average of once daily.
- ☐ 4 Falls more than once daily.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

10. Freezing when walking

- ☐ 0 None.
- ☐ 1 Rare freezing when walking; may have start-hesitation.
- ☐ 2 Occasional freezing when walking.
- ☐ 3 Frequent freezing. Occasionally falls from freezing.
- ☐ 4 Frequent falls from freezing.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

11. Walking

- ☐ 0 Normal.
- ☐ 1 Mild difficulty. May not swing arms or may tend to drag leg.
- ☐ 2 Moderate difficulty, but requires little or no assistance.
- ☐ 3 Severe disturbance of walking, requiring assistance.
- ☐ 4 Cannot walk at all, even with assistance.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

12. Tremor

- ☐ 0 Absent.
- ☐ 1 Slight and infrequently present.
- ☐ 2 Moderate; bothersome to participant.
- ☐ 3 Severe; interferes with many activities.
- ☐ 4 Marked; interferes with most activities.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

13. Sensory complaints related to parkinsonism

- ☐ 0 None.
- ☐ 1 Occasionally has numbness, tingling, or mild aching.
- ☐ 2 Frequently has numbness, tingling, or aching; not distressing.
- ☐ 3 Frequent painful sensations.
- ☐ 4 Excruciating pain.
- ☐ 8 Not applicable.
- ☐ 9 Unknown.

Form B3L: UPDRS Part III — Motor Examination[1]

This Form records the observations of the examiner for a single point in time. It is therefore limited, particularly for participants who have pronounced "ons" and "offs." Nevertheless, such information can be valuable, for it is objective, and the examiner may see the same participant over time in different "on" and "off" states that should be recorded.

The items **speech** and **gait** in this section are the objective scores obtained by the examiner listening and watching the participant walk, and therefore differ from the subjective ratings by the participant recorded in Form B2L. Gait may be affected only on one side, which is often the case in participants with hemiparkinsonism (Hoehn and Yahr Stage I).

In Form B2L, on ADL, **tremor** is graded subjectively by the participant. Many participants do not distinguish between resting tremor and action tremor. In Form B3L on motor examination, such a distinction is made. Therefore, there are separate items for **tremor at rest** and **action tremor**. Whereas both hands, both feet, and the face are all evaluated individually for resting tremor, only the arms are scored for action tremor. Action tremor elsewhere is much less frequent and would represent a sign of concurrent essential tremor. In contrast, action tremor of the arms in a participant with PD can be either an associated essential tremor or an action tremor due to PD (Lance et al., 1963).

Masked **facial expression** is a feature of bradykinesia. The definitions used for this item were enhanced compared to the original Columbia scale by making reference to the positioning of the lips. It is hoped that this guideline will serve to improve inter-rater reliability. **Rigidity**, like tremor at rest, is measured on five parts of the body.

Rapid succession movements of index finger, hand grip, forearm turning, and leg tapping are described, and these are important features of bradykinesia. These tests should be carried out for one extremity at a time, rather than right and left sides simultaneously. The **alternating pronation-supination** movements of the forearms can be carried out with the forearm horizontal or vertical to the ground. It should not be with the participant tapping the thighs, palms down alternating with palms up, since this maneuver adds another stimulus to the successive movements.

Arising from a chair is tested first by having the participant attempt to arise without the assistance of pushing off or pulling himself up by using his hands. If he/she is unable to succeed in standing, then he/she is allowed to use his hands as he/she wishes in order to arise. This is another measure of bradykinesia. The item **posture** scores a common dystonic feature of PD, whereas the item **postural stability** measures the righting reflex. The latter is tested by the "pull test," in which the examiner pulls on the participant's shoulders while standing behind him in order to catch him, should he/she start to fall. For accurate evaluation, the participant must be prepared by one or two practice trials of being pulled backward if he/she has not previously undergone the pull test. The item **body bradykinesia and hypokinesia** reflects total speed or slowness, as well as amplitude of movements. Since rapid successive movements of the limbs are rated in other items, this one refers specifically to slowness of movement of the entire body, particularly the trunk.

If it is not possible to evaluate a specific clinical symptom, indicate 8=Untestable and specify the reason.

Lance, J. W., Schwab, R. S., and Peterson, E. A. (1963): Action tremor and the cogwheel phenomenon in Parkinson's disease. Brain, 86:95-110.

Fahn S, Elton R. UPDRS Development Committee. The Unified Parkinson's Disease Rating Scale. In: Fahn S MC, Calne DB, Goldstein M, ed. Recent developments in Parkinson's disease. Vol 2. Florham Park, NJ: Macmillan Healthcare Information; 1987:153-163, 293-304.

[1]Fahn S, Elton RL, UPDRS Development Committee. The Unified Parkinson's Disease Rating Scale. In Fahn S, Marsden CD, Calne DB, Goldstein M, eds. Recent developments in Parkinson's disease, Vol. 2. Florham Park, NJ: Macmillan Healthcare Information, 1987:153-163, 293-304. Reproduced by permission of the author.

1. Speech

☐ 0 Normal.
☐ 1 Slight loss of expression, diction, and/or volume.
☐ 2 Monotone, slurred but understandable; moderately impaired.
☐ 3 Marked impairment, difficult to understand.
☐ 4 Unintelligible.
☐ 8 Untestable. (SPECIFY REASON): _____

2. Facial expression

☐ 0 Normal.
☐ 1 Minimal hypomimia, could be normal "poker face."
☐ 2 Slight but definitely abnormal diminution of facial expression.
☐ 3 Moderate hypomimia; lips parted some of the time.
☐ 4 Masked or fixed facies with severe or complete loss of facial expression; lips parted 1/4 inch or more.
☐ 8 Untestable. (SPECIFY REASON): _____

3. Tremor at rest

3a. Face, lips, chin

☐ 0 Absent.
☐ 1 Slight and infrequently present.
☐ 2 Mild in amplitude and persistent. Or moderate in amplitude, but only intermittently present.
☐ 3 Moderate in amplitude and present most of the time.
☐ 4 Marked in amplitude and present most of the time.
☐ 8 Untestable. (SPECIFY REASON): _____

3b. Right hand

☐ 0 Absent.
☐ 1 Slight and infrequently present.
☐ 2 Mild in amplitude and persistent. Or moderate in amplitude, but only intermittently present.
☐ 3 Moderate in amplitude and present most of the time.
☐ 4 Marked in amplitude and present most of the time.
☐ 8 Untestable. (SPECIFY REASON): _____

3c. Left hand

☐ 0 Absent.
☐ 1 Slight and infrequently present.
☐ 2 Mild in amplitude and persistent. Or moderate in amplitude, but only intermittently present.
☐ 3 Moderate in amplitude and present most of the time.
☐ 4 Marked in amplitude and present most of the time.
☐ 8 Untestable. (SPECIFY REASON): _____

3d. Right foot

☐ 0 Absent.

☐ 1 Slight and infrequently present.

☐ 2 Mild in amplitude and persistent. Or moderate in amplitude, but only intermittently present.

☐ 3 Moderate in amplitude and present most of the time.

☐ 4 Marked in amplitude and present most of the time.

☐ 8 Untestable. (SPECIFY REASON): _____

3e. Left foot

☐ 0 Absent.

☐ 1 Slight and infrequently present.

☐ 2 Mild in amplitude and persistent. Or moderate in amplitude, but only intermittently present.

☐ 3 Moderate in amplitude and present most of the time.

☐ 4 Marked in amplitude and present most of the time.

☐ 8 Untestable. (SPECIFY REASON): _____

4. Action or postural tremor of hands

4a. Right hand

☐ 0 Absent.

☐ 1 Slight; present with action.

☐ 2 Moderate in amplitude, present with action.

☐ 3 Moderate in amplitude with posture holding as well as action.

☐ 4 Marked in amplitude; interferes with feeding.

☐ 8 Untestable. (SPECIFY REASON): _____

4b. Left hand

☐ 0 Absent.

☐ 1 Slight; present with action.

☐ 2 Moderate in amplitude, present with action.

☐ 3 Moderate in amplitude with posture holding as well as action.

☐ 4 Marked in amplitude; interferes with feeding.

☐ 8 Untestable. (SPECIFY REASON): _____

5. Rigidity

(Judged on passive movement of major joints with participant relaxed in sitting position. Cogwheeling to be ignored.)

5a. Neck

☐ 0 Absent.
☐ 1 Slight or detectable only when activated by mirror or other movements.
☐ 2 Mild to moderate.
☐ 3 Marked, but full range of motion easily achieved.
☐ 4 Severe, range of motion achieved with difficulty.
☐ 8 Untestable. (SPECIFY REASON): _____

5b. Right upper extremity

☐ 0 Absent.
☐ 1 Slight or detectable only when activated by mirror or other movements.
☐ 2 Mild to moderate.
☐ 3 Marked, but full range of motion easily achieved.
☐ 4 Severe, range of motion achieved with difficulty.
☐ 8 Untestable. (SPECIFY REASON): _____

5c. Left upper extremity

☐ 0 Absent.
☐ 1 Slight or detectable only when activated by mirror or other movements.
☐ 2 Mild to moderate.
☐ 3 Marked, but full range of motion easily achieved.
☐ 4 Severe, range of motion achieved with difficulty.
☐ 8 Untestable. (SPECIFY REASON): _____

5d. Right lower extremity

☐ 0 Absent.
☐ 1 Slight or detectable only when activated by mirror or other movements.
☐ 2 Mild to moderate.
☐ 3 Marked, but full range of motion easily achieved.
☐ 4 Severe, range of motion achieved with difficulty.
☐ 8 Untestable. (SPECIFY REASON): _____

5e. Left lower extremity

☐ 0 Absent.
☐ 1 Slight or detectable only when activated by mirror or other movements.
☐ 2 Mild to moderate.
☐ 3 Marked, but full range of motion easily achieved.
☐ 4 Severe, range of motion achieved with difficulty.
☐ 8 Untestable. (SPECIFY REASON): _____

6. Finger taps

(Participant taps thumb with index finger in rapid succession with widest amplitude possible, each hand separately.)

6a. Right hand

☐ 0 Normal.

☐ 1 Mild slowing and/or reduction in amplitude.

☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.

☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.

☐ 4 Can barely perform the task.

☐ 8 Untestable. (SPECIFY REASON): _____

6b. Left hand

☐ 0 Normal.

☐ 1 Mild slowing and/or reduction in amplitude.

☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.

☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.

☐ 4 Can barely perform the task.

☐ 8 Untestable. (SPECIFY REASON): _____

7. Hand movements

(Participant opens and closes hands in rapid succession with widest amplitude possible, each hand separately.)

7a. Right hand

☐ 0 Normal.

☐ 1 Mild slowing and/or reduction in amplitude.

☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.

☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.

☐ 4 Can barely perform the task.

☐ 8 Untestable. (SPECIFY REASON): _____

7b. Left hand

☐ 0 Normal.

☐ 1 Mild slowing and/or reduction in amplitude.

☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.

☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.

☐ 4 Can barely perform the task.

☐ 8 Untestable. (SPECIFY REASON): _____

8. Rapid alternating movements of hands

(Pronation-supination movements of hands, vertically or horizontally, with as large an amplitude as possible, both hands simultaneously.)

8a. Right hand

- ☐ 0 Normal.
- ☐ 1 Mild slowing and/or reduction in amplitude.
- ☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.
- ☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.
- ☐ 4 Can barely perform the task.
- ☐ 8 Untestable. (SPECIFY REASON): _____

8b. Left hand

- ☐ 0 Normal.
- ☐ 1 Mild slowing and/or reduction in amplitude.
- ☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.
- ☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.
- ☐ 4 Can barely perform the task.
- ☐ 8 Untestable. (SPECIFY REASON): _____

9. Leg agility

(Participant taps heel on ground in rapid succession, picking up entire leg. Amplitude should be about 3 inches.)

9a. Right leg

- ☐ 0 Normal.
- ☐ 1 Mild slowing and/or reduction in amplitude.
- ☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.
- ☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.
- ☐ 4 Can barely perform the task.
- ☐ 8 Untestable. (SPECIFY REASON): _____

9b. Left leg

- ☐ 0 Normal.
- ☐ 1 Mild slowing and/or reduction in amplitude.
- ☐ 2 Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement.
- ☐ 3 Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movement.
- ☐ 4 Can barely perform the task.
- ☐ 8 Untestable. (SPECIFY REASON): _____

10. Arising from chair

(Participant attempts to arise from a straight-back wood or metal chair with arms folded across chest.)

- ☐ 0 Normal.
- ☐ 1 Slow; or may need more than one attempt.
- ☐ 2 Pushes self up from arms of seat.
- ☐ 3 Tends to fall back and may have to try more than one time, but can get up without help.
- ☐ 4 Unable to arise without help.
- ☐ 8 Untestable. (SPECIFY REASON): _____

11. Posture

- ☐ 0 Normal erect.
- ☐ 1 Not quite erect, slightly stooped posture; could be normal for older person.
- ☐ 2 Moderately stooped posture, definitely abnormal; can be slightly leaning to one side.
- ☐ 3 Severely stooped posture with kyphosis; can be moderately leaning to one side.
- ☐ 4 Marked flexion with extreme abnormality of posture.
- ☐ 8 Untestable. (SPECIFY REASON): _____

12. Gait

- ☐ 0 Normal.
- ☐ 1 Walks slowly, may shuffle with short steps, but no festination or propulsion.
- ☐ 2 Walks with difficulty, but requires little or no assistance; may have some festination, short steps, or propulsion.
- ☐ 3 Severe disturbance of gait, requiring assistance.
- ☐ 4 Cannot walk at all, even with assistance.
- ☐ 8 Untestable. (SPECIFY REASON): _____

13. Postural stability

(Response to sudden posterior displacement produced by pull on shoulders while participant erect with eyes open and feet slightly apart. Participant is prepared.)

- ☐ 0 Normal.
- ☐ 1 Retropulsion, but recovers unaided.
- ☐ 2 Absence of postural response; would fall if not caught by examiner.
- ☐ 3 Very unstable, tends to lose balance spontaneously.
- ☐ 4 Unable to stand without assistance.
- ☐ 8 Untestable. (SPECIFY REASON): _____

14. Body bradykinesia and hypokinesia

(Combining slowness, hesitancy, decreased armswing, small amplitude, and poverty of movement in general.)

- ☐ 0 None.
- ☐ 1 Minimal slowness, giving movement a deliberate character; could be normal for some persons. Possibly reduced amplitude.
- ☐ 2 Mild degree of slowness and poverty of movement which is definitely abnormal. Alternatively, some reduced amplitude.
- ☐ 3 Moderate slowness, poverty, or small amplitude of movement.
- ☐ 4 Marked slowness, poverty, or small amplitude of movement.
- ☐ 8 Untestable. (SPECIFY REASON): _____

15. Modified Hoehn and Yahr staging

- ☐ 0 Stage 0 = No signs of disease.
- ☐ 1 Stage 1 = Unilateral disease.
- ☐ 2 Stage 1.5 = Unilateral plus axial involvement.
- ☐ 3 Stage 2 = Bilateral disease, without impairment of balance.
- ☐ 4 Stage 2.5 = Mild bilateral disease, with recovery on pull test.
- ☐ 5 Stage 3 = Mild to moderate bilateral disease; some postural instability; physically independent.
- ☐ 6 Stage 4 = Severe disability; still able to walk or stand unassisted.
- ☐ 7 Stage 5 = Wheelchair-bound or bedridden unless aided.
- ☐ 8 Untestable. (SPECIFY REASON): _____

The staging of PD, as suggested by Hoehn and Yahr (1967), has been widely used, so its recording is provided within the UPDRS. Modifications to this staging have been suggested to provide more accuracy, and a modification of this staging scale has been incorporated into the UPDRS. Changes include a stage of 1.5 if there is hemiparkinsonism with axial involvement, such as neck rigidity, and a stage of 2.5 if there is recoverable postural instability. Stage 3 or greater is used for inability to recover on the "pull test." It should be noted that a participant with unilateral involvement of the dominant limbs could be more severely disabled than someone with mild bilateral disease. Thus, Stage 1 does not always accurately reflect a milder disorder than does Stage 2. Besides its insensitivity, the Hoehn and Yahr staging scale, therefore, is not always a reliable indicator of severity of parkinsonism.

If numerous clinical symptoms evaluated on this Form were untestable and thus it is not feasible to determine the Hoehn and Yahr stage, indicate 8=Untestable and specify the reason.

Hoehn, M. M., and Yahr, M. D. (1967): Parkinsonism: onset, progression and mortality. Neurology, 17:427-442.

Fahn S, Elton R. UPDRS Development Committee. The Unified Parkinson's Disease Rating Scale. In: Fahn S MC, Calne DB, Goldstein M, ed. Recent developments in Parkinson's disease. Vol 2. Florham Park, NJ: Macmillan Healthcare Information; 1987:153-163, 293-304.

Form B4L: Neuropsychiatric Inventory (NPI)[1]

Administration of the NPI

A. NPI interview

The NPI is based on responses from an informed caregiver, preferably one living with the participant.

The interview is best conducted with the caregiver in the absence of the participant to facilitate an open discussion of behaviors that may be difficult to describe with the participant present. Several points should be made when you introduce the NPI interview to the caregiver:

- Purpose of the interview
- Ratings — frequency, severity, distress (described below)
- Answers apply to behaviors that are new since the onset of the disease and have been present for the past four weeks
- Questions can usually be answered with "yes" or "no," and responses should be brief

When beginning the inventory, say to the caregiver, "These questions are designed to evaluate your [husband's/wife's/etc.] behavior. They can usually be answered 'yes' or 'no,' so please try to be brief in your responses." If the caregiver lapses into elaborate responses that provide little useful information, he/she may be reminded of the need to be brief. Some of the issues raised with this are very emotionally disturbing to caregivers, and the interviewer should reassure the caregiver that they will discuss the problems in more detail after completion of the inventory.

Questions should be asked exactly as written. Clarification should be provided if the caregiver does not understand the question. Acceptable clarifications are restatements of the questions in alternate terms.

B. Changes in behavior

The questions pertain to changes in the participant's behavior that have appeared since the onset of the illness. Behaviors that have been present throughout the participant's life and have not changed in the course of the illness are not scored even if they are abnormal (e.g., anxiety, depression). Behaviors are scored that have been present throughout life but have changed since the illness (e.g., the participant has always been apathetic but there has been a notable increase in apathy during the period of inquiry).

C. Screening questions

The screening question is asked to determine if the behavioral change is present or absent. If the answer to the screening question is negative, mark NO and proceed to the next screening question without asking the subquestions. If the answer to the screening question is positive or if there are any uncertainties in the caregiver's response or any inconsistencies between the response and other information known by the clinician (e.g., the caregiver responds negatively to the euphoria screening question but the participant appears euphoric to the clinician), the category is marked YES and is explored in more depth with the subquestions. If the subquestions confirm the screening question, the severity and frequency of the behavior are determined according to the criteria provided with each behavior.

In some cases, the caregiver will provide a positive response to the screening question and a negative reply to all subquestions. If this happens, ask the caregiver to expand on why he/she responded affirmatively to the screen. If he/she provides information relevant to the behavioral domain but in different terms, the behavior should be scored for severity and frequency as usual. If the original affirmative response was erroneous, leading to a failure to endorse any subquestions, then the behavior is changed to "NO" on the screen.

D. Frequency and severity ratings

When determining frequency and severity, use the behaviors identified by the subquestions as most aberrant. For example, if the caregiver indicates that resistive behavior is particularly problematic when you are asking the subquestions of the agitation section, then use resistive behavior to prompt judgments regarding the frequency and severity of agitation. If two behaviors are very problematic, use the frequency and severity of both behaviors to score the item. For example, if the participant has two or more types of delusions, then use the severity and the frequency of all delusional behaviors to phrase the questions regarding severity and frequency.

When determining frequency, say to the person being interviewed, "Now I want to find out how often these things [define using the description of the behaviors noted as most problematic on the subquestions] occur. Would you say that they occur less than once per week, about once per week, several times per week but not every day, or every day?" Some behaviors such as apathy eventually become continuously present, and then "are constantly present" can be substituted for "every day."

When determining severity, tell the person being interviewed, "Now I would like to find out how severe these behaviors are. By severity, I mean how disturbing or disabling they are for the participant. Would you say that [the behaviors] are mild, moderate, or severe?" Additional descriptors are provided in each section that may be used to help the interviewer clarify each grade of severity.

In each case, be sure that the caregiver provides you with a definite answer as to the frequency and severity of the behaviors. Do not guess what you think the caregiver would say based on your discussion.

It may be helpful to provide the caregiver with a piece of paper on which is written the frequency and severity descriptions (less than once per week, about once per week, several times per week and daily or continuously for frequency; and mild, moderate, and severe for severity) to allow him or her to visualize the response alternatives. This also saves the examiner from reiterating the alternatives with each question.

E. "Not applicable" designations

In very impaired participants or in participants with special medical circumstances, a set of questions may not be applicable. If the clinician or the caregiver believes that the questions are inappropriate, then enter 8=Not applicable for the first question of the section and record no further data for that section. Likewise, if the clinician feels that the responses are invalid (e.g., the caregiver did not seem to understand the particular set of questions asked), 8=Not applicable should also be marked.

Scoring the NPI

Frequency is rated as:

1 Occasionally — less than once per week
2 Often — about once per week
3 Frequently — several times per week but less than every day
4 Very frequently — daily or essentially continuously present

Severity is rated as:

1 Mild — produces little distress in the participant
2 Moderate — more disturbing to the participant but can be redirected by the caregiver
3 Severe — very disturbing to the participant and difficult to redirect

Distress is scored as:

0 — No distress
1 — Minimal
2 — Mild
3 — Moderate
4 — Moderately severe
5 — Very severe or extreme.

To calculate the score for each domain:
domain score = **frequency** × **severity**

Thus, for each behavioral domain there are **four scores:**

- Frequency
- Severity
- Total (frequency × severity)
- Caregiver distress

Inquire about symptoms the last four weeks before visit.

DELUSIONS

1. Does the participant have beliefs that you know are not true (for example, insisting that people are trying to harm him/her or steal from him/her)? Has he/she said that family members are not who they say they are or that the house is not their home? I'm not asking about mere suspiciousness; I am interested if the participant is convinced that these things are happening to him/her.

☐ 0 No **(SKIP TO QUESTION 2)**
☐ 1 Yes **(COMPLETE QUESTIONS 1a – 1i)**
☐ 8 Not applicable **(SKIP TO QUESTION 2)**

	1a.	Does the participant believe that he/she is in danger — that others are planning to hurt him/her?	☐ 0 No	☐ 1 Yes
	1b.	Does the participant believe that others are stealing from him/her?	☐ 0 No	☐ 1 Yes
	1c.	Does the participant believe that his/her spouse is having an affair?	☐ 0 No	☐ 1 Yes
	1d.	Does the participant believe that unwelcome guests are living in his/her house?	☐ 0 No	☐ 1 Yes
	1e.	Does the participant believe that his/her spouse or others are not who they claim to be?	☐ 0 No	☐ 1 Yes
	1f.	Does the participant believe that his/her house is not his/her home?	☐ 0 No	☐ 1 Yes
	1g.	Does the participant believe that family members plan to abandon him/her?	☐ 0 No	☐ 1 Yes
	1h.	Does the participant believe that television or magazine figures are actually present in the home? [Does he/she try to talk or interact with them?]	☐ 0 No	☐ 1 Yes
	1i.	Does the participant believe any other unusual things that I haven't asked about?	☐ 0 No	☐ 1 Yes

	1j.	If the screening question is confirmed, determine the frequency and severity of the delusions. FREQUENCY: ☐ 1 Occasionally — less than once per week ☐ 2 Often — about once per week ☐ 3 Frequently — several times per week but less than every day ☐ 4 Very frequently — once or more per day

	1k.	SEVERITY: ☐ 1 Mild — delusions present but seem harmless and produce little distress in the participant ☐ 2 Moderate — delusions are distressing and disruptive ☐ 3 Marked — delusions are very disruptive and are a major source of behavioral disruption (if PRN medications are prescribed, their use signals that the delusions are of marked severity)
	1l.	How emotionally distressing do you find this behavior? ☐ 0 Not at all ☐ 1 Minimally ☐ 2 Mildly ☐ 3 Moderately ☐ 4 Severely ☐ 5 Very severely or extremely

HALLUCINATIONS

2. Does the participant have hallucinations such as seeing false visions or hearing imaginary voices? Does he/she seem to see, hear, or experience things that are not present? By this question, we do not mean just mistaken beliefs such as stating that someone who has died is still alive; rather we are asking if the participant actually has abnormal experiences of sounds or visions.

☐ 0 No **(SKIP TO QUESTION 3)**
☐ 1 Yes **(COMPLETE QUESTIONS 2a – 2j)**
☐ 8 Not applicable **(SKIP TO QUESTION 3)**

	2a.	Does the participant describe hearing voices or acts if he/she hears voices?	☐ 0 No	☐ 1 Yes
	2b.	Does the participant talk to people who are not there?	☐ 0 No	☐ 1 Yes
	2c.	Does the participant describe seeing things not seen by others or behave as if he/she is seeing things not seen by others?	☐ 0 No	☐ 1 Yes
	2d.	Does the participant report smelling odors not smelled by others?	☐ 0 No	☐ 1 Yes
	2e.	Does the participant describe feeling things on his/her skin or otherwise appear to be feeling things crawling or touching him/her?	☐ 0 No	☐ 1 Yes
	2f.	Does the participant describe tastes that are without any known cause?	☐ 0 No	☐ 1 Yes
	2g.	Does the participant describe any other unusual sensory experiences?	☐ 0 No	☐ 1 Yes

	2h.	If the screening question is confirmed, determine the frequency and severity of the hallucinations. FREQUENCY: ☐ 1 Occasionally — less than once per week ☐ 2 Often — about once per week ☐ 3 Frequently — several times per week but less than every day ☐ 4 Very frequently — once or more per day

	2i.	SEVERITY: ☐ 1 Mild — hallucinations are present but seem harmless and cause little distress for the participant ☐ 2 Moderate — hallucinations are distressing and are disruptive to the participant ☐ 3 Marked — hallucinations are very disruptive and are a major source of behavioral disturbance. PRN medications may be required to control them.		
	2j.	How emotionally distressing do you find this behavior? ☐ 0 Not at all ☐ 1 Minimally ☐ 2 Mildly ☐ 3 Moderately ☐ 4 Severely ☐ 5 Very severely or extremely		
ANXIETY				
3.	**Is the participant very nervous, worried, or frightened for no apparent reason? Does he/she seem very tense or fidgety? Is the participant afraid to be apart from you?** ☐ 0 No **(SKIP TO QUESTION 4)** ☐ 1 Yes **(COMPLETE QUESTIONS 3a – 3j)** ☐ 8 Not applicable **(SKIP TO QUESTION 4)**			
	3a.	Does the participant say that he/she is worried about planned events?	☐ 0 No	☐ 1 Yes
	3b.	Does the participant have periods of feeling shaky, unable to relax, or feeling excessively tense?	☐ 0 No	☐ 1 Yes
	3c.	Does the participant have periods of (or complain of) shortness of breath, gasping, or sighing for no apparent reason other than nervousness?	☐ 0 No	☐ 1 Yes
	3d.	Does the participant complain of butterflies in his/her stomach, or of racing or pounding of the heart in association with nervousness (symptoms not explained by ill health)?	☐ 0 No	☐ 1 Yes
	3e.	Does the participant avoid certain places or situations that make him/her more nervous such as riding in the car, meeting with friends, or being in crowds?	☐ 0 No	☐ 1 Yes
	3f.	Does the participant become nervous and upset when separated from you [or his/her caregiver]? [Does he/she cling to you to keep from being separated?]	☐ 0 No	☐ 1 Yes
	3g.	Does the participant show any other signs of anxiety?	☐ 0 No	☐ 1 Yes
	3h.	If the screening question is confirmed, determine the frequency and severity of the anxiety. FREQUENCY: ☐ 1 Occasionally — less than once per week ☐ 2 Often — about once per week ☐ 3 Frequently — several times per week but less than every day ☐ 4 Very frequently — once or more per day		

	3i.	SEVERITY: ☐ 1 Mild — anxiety is distressing but usually responds to redirection or reassurance ☐ 2 Moderate — anxiety is distressing, anxiety symptoms are spontaneously voiced by the participant and difficult to alleviate ☐ 3 Marked — anxiety is very distressing and a major source of suffering for the participant
	3j.	How emotionally distressing do you find this behavior? ☐ 0 Not at all ☐ 1 Minimally ☐ 2 Mildly ☐ 3 Moderately ☐ 4 Severely ☐ 5 Very severely or extremely

APATHY / INDIFFERENCE

4. Has the participant lost interest in the world around him/her? Has he/she lost interest in doing things or lack motivation for starting new activities? Is he/she more difficult to engage in conversation or in doing chores? Is the participant apathetic or indifferent?

☐ 0 No **(SKIP TO QUESTION 5)**
☐ 1 Yes **(COMPLETE QUESTIONS 4a – 4k)**
☐ 8 Not applicable **(SKIP TO QUESTION 5)**

	4a.	Does the participant seem less spontaneous and less active than usual?	☐ 0 No	☐ 1 Yes
	4b.	Is the participant less likely to initiate a conversation?	☐ 0 No	☐ 1 Yes
	4c.	Is the participant less affectionate or lacking in emotions when compared to his/her usual self?	☐ 0 No	☐ 1 Yes
	4d.	Does the participant contribute less to household chores?	☐ 0 No	☐ 1 Yes
	4e.	Does the participant seem less interested in the activities and plans of others?	☐ 0 No	☐ 1 Yes
	4f.	Has the participant lost interest in friends and family members?	☐ 0 No	☐ 1 Yes
	4g.	Is the participant less enthusiastic about his/her usual interests?	☐ 0 No	☐ 1 Yes
	4h.	Does the participant show any other signs that he/she doesn't care about doing new things?	☐ 0 No	☐ 1 Yes

	4i.	If the screening question is confirmed, determine the frequency and severity of the apathy/indifference. FREQUENCY: ☐ 1 Occasionally — less than once per week ☐ 2 Often — about once per week ☐ 3 Frequently — several times per week but less than every day ☐ 4 Very frequently — nearly always present

4j. SEVERITY:

☐ 1 Mild — apathy is notable but produces little interference with daily routines; only mildly different from participant's usual behavior; participant responds to suggestions to engage in activities

☐ 2 Moderate — apathy is very evident; may be overcome by the caregiver with coaxing and encouragement; responds spontaeously only to powerful events such as visits from close relatives or family members

☐ 3 Marked — apathy is very evident and usually fails to respond to any encouragement or external events

4k. How emotionally distressing do you find this behavior?

☐ 0 Not at all

☐ 1 Minimally

☐ 2 Mildly

☐ 3 Moderately

☐ 4 Severely

☐ 5 Very severely or extremely

SUPPLEMENTAL INFORMATION*

**Items are not part of NPI*

For all questions related to medication use, determine the drugID by using the **drugID LookUp Tool** on the NACC website at http://www.alz.washington.edu/MEMBER/DrugCodeLookUp.html

5. Is the participant currently on **dopaminergic agents**?

☐ 0 No **(SKIP TO QUESTION 6)**

☐ 1 Yes **(CONTINUE TO QUESTION 5a)**

☐ 9 Unknown **(SKIP TO QUESTION 6)**

5a. Age at initiation of dopaminergic agents:

Questions 5a1 – 5a6, below: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of first dopaminergic use. For example, if the co-participant indicates that use began in the participant's 50s, an estimated age of 55 years could be provided.

If the dose cannot be ascertained, write in "Unknown."

Age *(999=unknown)*	**Drug code (drugID)**	**Dose**
5a1. ___ ___ ___	5a2. d ___ ___ ___ ___ ___	5a3. _____
If not applicable, leave 5a4 – 5a6 blank:		
5a4. ___ ___ ___	5a5. d ___ ___ ___ ___ ___	5a6. _____

6. If the participant had no delusions, hallucinations, anxiety, or apathy reported in Questions 1 – 4, **END FORM HERE**. Otherwise, if the participant has delusions (Question 1 is 1=Yes), then **ANSWER QUESTIONS 6a AND 6b**. If the participant does not have delusions or if the question is not applicable (Question 1 is 0=No or 8=Not applicable), then enter 888=Not applicable for Question 6a and **SKIP TO QUESTION 7**.

6a. Age of onset of delusions: ___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 6a: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of delusions. For example, if the co-participant indicates that onset occurred in the participant's 50s, an estimated age of 55 years could be provided.

6b. Delusions currently being treated with medication?
- ☐ 0 No **(SKIP TO QUESTION 7)**
- ☐ 1 Yes **(CONTINUE TO QUESTION 6c – 6d)**
- ☐ 9 Unknown **(SKIP TO QUESTION 7)**

6c. Medication 1: d ___ ___ ___ ___ ___

6d. Medication 2: d ___ ___ ___ ___ ___
Leave blank if not applicable

7. If the participant has hallucinations (Question 2 is 1=Yes), then **ANSWER QUESTIONS 7a AND 7b**. If the participant does not have hallucinations or if the question is not applicable (Question 2 is 0=No or 8=Not applicable), then enter 888=Not applicable for Question 7a and **SKIP TO QUESTION 8**.

7a. Age of onset of hallucations: ___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 7a: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of hallucinations. For example, if the co-participant indicates that onset occurred in the participant's 50s, an estimated age of 55 years could be provided.

7b. Hallucinations currently being treated with medication?
- ☐ 0 No **(SKIP TO QUESTION 8)**
- ☐ 1 Yes **(CONTINUE TO QUESTION 7c – 7d)**
- ☐ 9 Unknown **(SKIP TO QUESTION 8)**

7c. Medication 1: d ___ ___ ___ ___ ___

7d. Medication 2: d ___ ___ ___ ___ ___
Leave blank if not applicable

8. If the participant has anxiety (Question 3 is 1=Yes), then **ANSWER QUESTIONS 8a AND 8b**. If the participant does not have anxiety or if the question is not applicable (Question 3 is 0=No or 8=Not applicable), then enter 888=Not applicable for Question 8a and **SKIP TO QUESTION 9**.

8a. Age of onset of anxiety: ___ ___ ___ *(888=Not applicable; 999=Unknown)*

Question 8a: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of anxiety. For example, if the co-participant indicates that onset occurred in the participant's 50s, an estimated age of 55 years could be provided.

8b. Anxiety currently being treated with medication?
- ☐ 0 No **(SKIP TO QUESTION 9)**
- ☐ 1 Yes **(CONTINUE TO QUESTION 8c – 8d)**
- ☐ 9 Unknown **(SKIP TO QUESTION 9)**

	8c. Medication 1: d ___ ___ ___ ___ ___	8d. Medication 2: d ___ ___ ___ ___ ___ *Leave blank if not applicable*

9. If the participant is apathetic or indifferent (Question 4 is 1=Yes), then **ANSWER QUESTIONS 9a AND 9b**. If the participant is not apathetic or indifferent, or if the question is not applicable (Question 4 is 0=No or 8=Not applicable), then enter 888=Not applicable for Question 9a and **END FORM HERE**.

9a. Age of onset of apathy/indifference: ___ ___ ___ *(888=Not applicable; 999= Unknown)*

Question 9a: If the exact age is unknown, the clinician must use his/her best judgment to estimate the age of onset of apathy. For example, if the co-participant indicates that onset occurred in the participant's 50s, an estimated age of 55 years could be provided.

9b. Apathy/indifference currently being treated with medication?

☐ 0 No **(END FORM HERE)**
☐ 1 Yes **(CONTINUE TO QUESTION 9c – 9d)**
☐ 9 Unknown **(END FORM HERE)**

9c. Medication 1: d ___ ___ ___ ___ ___	9d. Medication 2: d ___ ___ ___ ___ ___ *Leave blank if not applicable*

References

Cummings JL, Mega M, Gray K, Rosenberg-Thompson S, Carusi DA, Gornbein J. The Neuropsychiatric Inventory: comprehensive assessment of psychopathology in dementia. Neurology 1994; 44: 2308-2314.

Cummings JL. The Neuropsychiatric Inventory: Assessing psychopathology in dementia patients. Neurology 1997; 48 (Suppl. 6): S10-S16.

Kaufer DI, Cummings JL, Christine D, Bray T, Castellon S, Masterman D, MacMillan A, Ketchel P, Dekosky ST. Assessing the impact of neuropsychiatric symptoms in Alzheimer's disease: the Neuropsychiatric Inventory Caregiver Distress Scale. J Am Geriatric Soc 1998; 46: 210-215.

Wood S, Cummings JL, Hsu M-A, Barclay T, Wheatley MV, Yarema KT, Schnelle JF. The use of the Neuropsychiatric Inventory in nursing home residents: characterization and measurement. Am J Geriatr Psychiatry 1999;8:75-83.

Form B5L: Mayo Fluctuations Scale

Questions 1 – 4: The four questions included in the Mayo Fluctuations Scale have been found to significantly differentiate AD from DLB. Positive items are summed to create a "fluctuations composite score," with a score of 3 or 4 previously associated with a positive predictive value of 83% for distinguishing those with clinical DLB from those with AD (Ferman et al., 2004).

Reference:

T. J. Ferman, G. E. Smith, B. F. Boeve, R. J. Ivnik, R. C. Petersen, D. Knopman, N. Graff-Radford, J. Parisi, and D. W. Dickson. DLB fluctuations: Specific features that reliably differentiate DLB from AD and normal aging. Neurology;62(2):181-187.

DIRECTIONS: Please mark the answer that best describes the participant within the past 6 months.	
1. **Is the participant drowsy and lethargic during the day, despite getting enough sleep the night before?**	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
2. **Does the participant sleep 2 or more hours during the day (before 7:00 p.m.)?**	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
3. **Are there times when the participant's flow of ideas is disorganized, unclear, or not logical?**	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
4. **Does the participant tend to stare into space for long periods of time?**	☐ 0 No ☐ 1 Yes ☐ 9 Unknown

Form B6L: Mayo Sleep Questionnaire — Participant

FOR CLINICIAN USE ONLY

0. Is the participant too cognitively impaired (e.g., CDR>1) to complete this form?

☐ 0 No **(CONTINUE TO ADMINISTER QUESTIONNAIRE)**

☐ 1 Yes **(END FORM HERE)**

For details on the sensitivity and specificity of detecting RBD, please refer to:

Bradley F. Boeve, Jennifer R. Molano, Tanis J. Ferman, Glenn E. Smith, Siong-Chi Lin, Kevin Bieniek, Wael Haidar, Maja Tippmann-Peikert, David S. Knopman, Neill R. Graff-Radford, John A. Lucas, Ronald C. Petersen, and Michael H. Silber. Validation of the Mayo Sleep Questionnaire to Screen for REM Sleep Behavior Disorder in an Aging and Dementia Cohort. Sleep Med. 2011 May; 12(5): 445–453.

Please mark "Yes" if the described event has occurred at least 3 times.

1. **Have you ever been told that you seem to "act out your dreams" while sleeping (punched or flailed arms in the air, shouted or screamed)?**	☐ 0 No **(SKIP TO QUESTION 2)** ☐ 1 Yes **(COMPLETE QUESTIONS 1a – 1e)**
1a. **How many months or years has this been going on?**	___ ___ year(s) ___ ___ month(s)
If the participant has experienced this symptom for less than one year, enter "0" for year(s) and the number of months. If the participant has experienced this symptom for more than one year, enter the number of full years in the year(s) section, and enter any partial years in the month(s) section. If reporting only years, enter "0" in the month(s) section.	
1b. **Have you ever been injured from these behavior (bruises, cuts, broken bones)?**	☐ 0 No ☐ 1 Yes
1c. **Has a bedpartner ever been injured from these behaviors (bruises, blows, pulled hair)?**	☐ 0 No ☐ 1 Yes ☐ 8 No bedpartner

1d. **Have you had dreams of being chased or attacked, or that involve defending yourself?**	☐ 0 No ☐ 1 Yes
1e. **Have you been told that you make movements while sleeping that seem to match the details of your dream?**	☐ 0 No ☐ 1 Yes
2. **Have you been told that your legs repeatedly jerk or twitch during sleep (not just when falling asleep)?**	☐ 0 No ☐ 1 Yes
3. **Does a restless, nervous, tingly, or creepy-crawly feeling in your legs make it hard to fall or stay asleep?**	☐ 0 No **(SKIP TO QUESTION 4)** ☐ 1 Yes **(COMPLETE QUESTIONS 3a – 3c)**
3a. **Do you experience an irresistible urge to move the legs at those times?**	☐ 0 No ☐ 1 Yes
3b. **Do the uncomfortable leg sensations decrease when you move them or when you walk around?**	☐ 0 No ☐ 1 Yes
3c. **When do these sensations seem to be worse?**	☐ 1 Before 6:00 p.m. ☐ 2 After 6:00 p.m.
4. **Have you ever walked around the bedroom or house in your sleep?**	☐ 0 No ☐ 1 Yes
5. **Have you ever snorted or choked yourself awake?**	☐ 0 No ☐ 1 Yes
6. **Have you ever been told that you stop breathing in your sleep?**	☐ 0 No **(SKIP TO QUESTION 7)** ☐ 1 Yes **(COMPLETE QUESTION 6a)**
6a. **Are you currently being treated for this (e.g., CPAP)?**	☐ 0 No ☐ 1 Yes
7. **Do you experience leg cramps at night (e.g., also called a "charlie horse" with intense pain in certain muscles in the leg)?**	☐ 0 No ☐ 1 Yes
8. **Rate your general level of alertness for the past 3 weeks on a scale from 0 to 10:** ___ ___ 0——1——2——3——4——5——6——7——8——9——10 Sleep all day — Fully and normally awake	

Form B7L: Mayo Sleep Questionnaire — Co-participant

For details on the sensitivity and specificity of detecting RBD, please refer to:

Bradley F. Boeve, Jennifer R. Molano, Tanis J. Ferman, Glenn E. Smith, Siong-Chi Lin, Kevin Bieniek, Wael Haidar, Maja Tippmann-Peikert, David S. Knopman, Neill R. Graff-Radford, John A. Lucas, Ronald C. Petersen, and Michael H. Silber. Validation of the Mayo Sleep Questionnaire to Screen for REM Sleep Behavior Disorder in an Aging and Dementia Cohort. Sleep Med. 2011 May; 12(5): 445–453.

1. **Do you live with the participant?**	☐ 0 No **(END FORM HERE)** ☐ 1 Yes **(CONTINUE TO QUESTION 2)**
2. **Do you sleep in the same room as the participant?**	☐ 0 No **(CONTINUE TO QUESTION 2a)** ☐ 1 Yes **(SKIP TO QUESTION 3)**
2a. **If no, is it because of his/her sleep behaviors (i.e., snores too loud, acts out dreams, etc.)?**	☐ 0 No ☐ 1 Yes

Please mark "Yes" if the described event has occurred <u>at least 3 times</u>.

3.	**Have you ever seen the participant appear to "act out his/her dreams" while sleeping (punched or flailed arms in the air, shouted, or screamed)?**		☐ 0 No **(SKIP TO QUESTION 4)** ☐ 1 Yes **(COMPLETE QUESTIONS 3a – 3e)**
	3a.	**How many months or years has this been going on?**	___ ___ year(s) ___ ___ month(s)
If the participant has experienced this symptom for less than one year, enter "0" for year(s) and the number of months. If the participant has experienced this symptom for more than one year, enter the number of full years in the year(s) section, and enter any partial years in the month(s) section. If reporting only years, enter "0" in the month(s) section.			
	3b.	**Has the participant ever been injured from these behaviors (bruises, cuts, broken bones)?**	☐ 0 No ☐ 1 Yes
	3c.	**Has a bedpartner ever been injured from these behaviors (bruises, blows, pulled hair)?**	☐ 0 No ☐ 1 Yes ☐ 8 No bedpartner

	3d.	**Has the participant told you about dreams of being chased or attacked, or that involve defending himself/herself?**	☐ 0 No ☐ 1 Yes ☐ 8 Never told me about dreams
	3e.	**If the participant woke up and told you about a dream, did the details of the dream match the movements made while sleeping?**	☐ 0 No ☐ 1 Yes ☐ 8 Never told me about dreams
4.		**Do the participant's legs repeatedly jerk or twitch during sleep (not just when falling asleep)?**	☐ 0 No ☐ 1 Yes
5.		**Does the participant complain of a restless, nervous, tingly, or creepy-crawly feeling in his/her legs that disrupts his/her ability to fall or stay asleep?**	☐ 0 No **(SKIP TO QUESTION 6)** ☐ 1 Yes **(COMPLETE QUESTIONS 5a – 5b)**
	5a.	**Does the participant tell you that these leg sensations decrease when he/she moves them or walks around?**	☐ 0 No ☐ 1 Yes
	5b.	**When do these sensations seem to be the worst?**	☐ 1 Before 6:00 p.m. ☐ 2 After 6:00 p.m.
6.		**Has the participant ever walked around the bedroom or house while asleep?**	☐ 0 No ☐ 1 Yes
7.		**Has the participant ever snorted or choked him/herself awake?**	☐ 0 No ☐ 1 Yes
8.		**Does the participant ever seem to stop breathing during sleep?**	☐ 0 No **(SKIP TO QUESTION 9)** ☐ 1 Yes **(COMPLETE QUESTION 8a)**
	8a.	**Is the participant currently being treated for this (e.g., CPAP)?**	☐ 0 No ☐ 1 Yes
9.		**Does the participant have leg cramps at night (e.g., also called a "charlie horse" with intense pain in certain muscles in the leg)?**	☐ 0 No ☐ 1 Yes
10.		**Rate the participant's general level of alertness for the past 3 weeks on a scale from 0 to 10:** ___ ___ 0——1——2——3——4——5——6——7——8——9——10 Sleep all day — Fully and normally awake	

Form B8L: SCOPA Sleep — Participant

Details on the reliability and validity of the SCOPA Sleep instrument can be found in:

Johan Marinus; Martine Visser; Jacobus Johannes van Hilten; Gert Jan Lammers; Anne Margarethe Stiggelbout. Assessment of Sleep and Sleepiness in Parkinson Disease. SLEEP 2003;26(8):1049-54.

FOR CLINICIAN USE ONLY

0. Is the participant too cognitively impaired (e.g., CDR>1) to complete this form?

☐ 0 No **(CONTINUE TO ADMINISTER QUESTIONNAIRE)**

☐ 1 Yes **(END FORM HERE)**

PARTICIPANT INSTRUCTIONS

By means of this questionnaire, we would like to find out to what extent *in the past month* you have had problems with sleeping. Some of the questions are about problems with sleeping *at night*, such as, for example, not being able to fall asleep or not managing to sleep on. Another set of questions is about problems with sleeping *during the day*, such as dozing off (too) easily and having trouble staying awake.

First read these instructions before you answer the questions!

Place a cross in the box corresponding to the answer that best reflects your situation. If you wish to change an answer, fill in the "wrong" box and place a cross in the correct one. If you have been using sleeping tablets, then the answer should reflect how you have slept while taking these tablets.

Nighttime sleep

In the past month, how often have you …	Not at all	A little	Quite a bit	A lot
1. Had trouble falling asleep when you went to bed at night	☐ 0	☐ 1	☐ 2	☐ 3
2. Felt that you have woken too often	☐ 0	☐ 1	☐ 2	☐ 3
3. Felt that you have been lying awake for too long at night	☐ 0	☐ 1	☐ 2	☐ 3
4. Felt that you have woken too early in the morning	☐ 0	☐ 1	☐ 2	☐ 3
5. Felt you have had too little sleep at night	☐ 0	☐ 1	☐ 2	☐ 3

Adapted from Marinus J, Visser M, van Hilten JJ, Lammers GJ, Stiggelbout AM. Assessment of sleep and sleepiness in Parkinson disease. SLEEP 2003;26:1049-1054. For further information, please contact Dr. J. Marinus, Leiden University Medical Center, Department of Neurology (K5Q), P.O. Box 9600, NL-2300 RC Leiden (email: j.marinus@lumc.nl).

Nighttime sleep, cont.

6. Overall, how well have you slept at night during the past month? (CHOOSE ONE):

- ☐ 1 Very well
- ☐ 2 Well
- ☐ 3 Rather well
- ☐ 4 Not well but not badly
- ☐ 5 Rather badly
- ☐ 6 Badly
- ☐ 7 Very badly

Daytime sleepiness

In the past month, how often have you …	Never	Sometimes	Regularly	Often
7. Fallen asleep unexpectedly during the day or in the evening	☐ 0	☐ 1	☐ 2	☐ 3
8. Fallen asleep while sitting peacefully	☐ 0	☐ 1	☐ 2	☐ 3
9. Fallen asleep while watching TV or reading	☐ 0	☐ 1	☐ 2	☐ 3
10. Fallen asleep while talking to someone	☐ 0	☐ 1	☐ 2	☐ 3
11. Had trouble staying awake during the day or in the evening	☐ 0	☐ 1	☐ 2	☐ 3
12. Experienced falling asleep during the day as a problem	☐ 0	☐ 1	☐ 2	☐ 3

Form B9L: SCOPA Sleep — Co-participant

Details on the reliability and validity of the SCOPA Sleep instrument can be found in:

Johan Marinus; Martine Visser; Jacobus Johannes van Hilten; Gert Jan Lammers; Anne Margarethe Stiggelbout. Assessment of Sleep and Sleepiness in Parkinson Disease. SLEEP 2003;26(8):1049-54.

Section I: Co-participant

CO-PARTICIPANT INSTRUCTIONS

By means of this questionnaire, we would like to find out to what extent *in the past month* the participant has had problems with sleeping. Some of the questions are about problems with sleeping *at night*, such as, for example, not being able to fall asleep or not managing to sleep on. Another set of questions is about problems with sleeping *during the day*, such as dozing off (too) easily and having trouble staying awake.

First read these instructions before you answer the questions!

Place a cross in the box corresponding to the answer that best reflects the situation. If you wish to change an answer, fill in the "wrong" box and place a cross in the correct one. If the participant has been using sleeping tablets, then the answer should reflect how s/he has slept while taking these tablets.

Nighttime sleep

In the past month, how often has the participant …	Not at all	A little	Quite a bit	A lot
1. Had trouble falling asleep when they went to bed at night	☐ 0	☐ 1	☐ 2	☐ 3

Questions 2-5: Responses should be based on the participant's nighttime sleep that the co-participant has observed in the past month and, if known, what the participant has expressed about their nighttime sleep in the past month.

2. Felt that they have woken too often	☐ 0	☐ 1	☐ 2	☐ 3
3. Felt that they been lying awake for too long at night	☐ 0	☐ 1	☐ 2	☐ 3
4. Felt that they have woken too early in the morning	☐ 0	☐ 1	☐ 2	☐ 3
5. Felt they have had too little sleep at night	☐ 0	☐ 1	☐ 2	☐ 3

☐1 Very well
☐2 Well
☐3 Rather well
☐4 Not well but not badly
☐5 Rather badly
☐6 Badly
☐7 Very badly

Adapted from Marinus J, Visser M, van Hilten JJ, Lammers GJ, Stiggelbout AM. Assessment of sleep and sleepiness in Parkinson disease. SLEEP 2003;26:1049-1054. For further information, please contact Dr. J. Marinus, Leiden University Medical Center, Department of Neurology (K5Q), P.O. Box 9600, NL-2300 RC Leiden (email: j.marinus@lumc.nl).

Daytime sleepiness				
In the past month, how often has the participant …	**Never**	**Sometimes**	**Regularly**	**Often**
7. Fallen asleep unexpectedly during the day or in the evening	☐ 0	☐ 1	☐ 2	☐ 3
8. Fallen asleep while sitting peacefully	☐ 0	☐ 1	☐ 2	☐ 3
9. Fallen asleep while watching TV or reading	☐ 0	☐ 1	☐ 2	☐ 3
10. Fallen asleep while talking to someone	☐ 0	☐ 1	☐ 2	☐ 3
11. Had trouble staying awake during the day or in the evening	☐ 0	☐ 1	☐ 2	☐ 3
12. Experienced falling asleep during the day as a problem	☐ 0	☐ 1	☐ 2	☐ 3

Section II: Clinician

First predominant symptom

13. Indicate the first predominant symptom to appear during the participant's lifetime.
☐ 1 Disturbed nighttime sleep
☐ 2 Excessive daytime sleepiness
☐ 8 Not applicable — never experienced disturbed nighttime sleep or excessive daytime sleepiness
If not applicable, **SKIP TO QUESTION 16**

14. At what age did the disturbed nighttime sleep first appear? ___ ___ ___ *(888=Not applicable; 999=Unknown)*

15. At what age did the excessive daytime sleepiness first appear? ___ ___ ___ *(888=Not applicable; 999=Unknown)*

16. WAS A STANDARDIZED SCALE OF DAYTIME SLEEPINESS COMPLETED AT THIS VISIT?
☐ 0 No **(END FORM HERE)** ☐ 1 Yes **(CONTINUE TO QUESTIONS 16a and 16b)**

16a. Which version?
☐ 1 Epworth
☐ 2 Stanford
☐ 3 Other (SPECIFY): _____

16b. What was the score? _____ *(999 = Unknown)*

Form C1L: Neuropsychological Battery Scores

This form should be completed by Alzheimer's Disease Center or clinic staff, based on participant response. If the participant cannot complete a particular test, refer to the appropriate key for coding entry.

The instructions provided within the LBD Module—Worksheets For Tests Reported on Form C1L should be closely followed at all times since these instructions may be different from Center-specific protocols that may already be in place.

Some participants may self-correct during the course of performance after an initial erroneous response. If this occurs, count the self-corrected response only if it occurs immediately after the error has been made.

Part 1: Speeded Attention Task (below)

Performance in the Speeded Attention Task measures processing speed and executive functioning by evaluating attention with and without conflicting cues. In this test, participants are asked to complete three related tasks as quickly as possible: (1) reading a list of words on a page, (2) naming the colors of printed symbols, and (3) naming the color of the ink, while ignoring the printed word. The measures of performance are the total numbers of correctly read words or named colors.

Review the instructions in the LBD Module — Worksheets for Tests Reported on Form C1L, complete the worksheet, and enter the appropriate score for each task. If the test could not be completed, enter the appropriate reason code.

1. Speeded Attention Task *If test not completed, enter reason code (995 – 998) for Question 1a, and* **SKIP TO QUESTION 2**	
1a. Raw Word Score:	___ ___ ___ *(0 – 150, 995 – 998)*
1b. Raw Color Score:	___ ___ ___ *(0 – 150)*
1c. Raw Color-word score:	___ ___ ___ *(0 – 150)*

Part 2: Noise Pareidolia Task (below)

The Noise Pareidolia Task is a test of visual perceptual processing. The task involves visual discrimination and measures visual misperceptions or visual illusions (pareidolia) that can be evoked by ambiguous stimuli. Participants are presented with black and white patterns, some of which contain images of human faces within the pattern, and others do not. If a face is detected by the participant, they are asked to respond "Yes" and point to the face. If a face is not detected, the participant is asked to respond "No." The primary measures are the total correct responses and the number of illusory noise (pareidolia) responses (items in which a face was identified by the participant, but the pattern was without a face image).

Review the instructions in the LBD Module—Worksheets for Tests Reported on Form C1L, complete the worksheet, and enter the appropriate score for each task. If the test could not be completed, enter the appropriate reason code.

2. Noise Pareidolia Task *If test not completed, enter reason code (95 – 98) for Question 2a, and* **END FORM HERE**	
2a. Correct **YES** Face Responses:	___ ___ *(0 – 7, 95 – 98)*
2b. Correct **NO** Noise Responses:	___ ___ *(0 – 13)*
2c. Total **YES** and **NO** Correct:	___ ___ *(0 – 20)*
2d. Pareidolia (Illusory) Responses:	___ ___ *(0 – 13)*

Form E1L: Genetics

Questions 1–7, DLB and PDD mutations

If the participant has evidence of a LRRK2, PARK2, PARK7, PINK1, SNCA, or GBA mutation, select 1=Yes for the question corresponding to the specific mutation. If medical record review and/or testing has been done on the participant and he/she has no LRRK2, PARK2, PARK7, PINK1, SNCA, or GBA mutation, select 0=No for the question corresponding to the specific mutation. If sufficient evidence is not available (e.g., no testing done), select 9=Unknown.

If the participant has evidence of another DLB or PDD mutation, beyond what is captured in Questions 1-6, enter the mutation in the Other (SPECIFY) field, and indicate whether the findings were positive (1=Yes) or negative (0=No). If no evidence of another DLB or PDD mutation is available, select 9=Unknown.

For each mutation, indicate the information source. If laboratory documentation is available, select 1=Commercial laboratory test documentation if the test was completed by a commercial source, or 2=Research laboratory test documentation if the test was completed by a healthcare or research source. If no laboratory test documentation is available and the mutation is reported by the family, select 3=Family report. If the information source is unknown, select 9=Unknown.

Does the participant have any of the following mutations (select only one answer per question):

Mutation	No	Yes	Unknown	If yes, information source (see KEY)
1. LRRK2	☐ 0	☐ 1	☐ 9	1a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
2. PARK2	☐ 0	☐ 1	☐ 9	2a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
3. PARK7	☐ 0	☐ 1	☐ 9	3a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
4. PINK1	☐ 0	☐ 1	☐ 9	4a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
5. SNCA	☐ 0	☐ 1	☐ 9	5a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
6. GBA	☐ 0	☐ 1	☐ 9	6a. ☐ 1 ☐ 2 ☐ 3 ☐ 9
7. Other (SPECIFY): _____	☐ 0	☐ 1	☐ 9	7a. ☐ 1 ☐ 2 ☐ 3 ☐ 9

KEY: 1 = Commercial laboratory test documentation
2 = Research laboratory test documentation
3 = Family report (select only if no laboratory test was done)
9 = Unknown

Form E2L: Neuroimaging Available and Findings

STRUCTURAL MRI	
1. Has the participant had at least one structrual MRI scan, obtained as part of the current evaluation or a previous evaluation?	☐ 0 **No or unknown (SKIP TO QUESTION 2)** ☐ 1 **Yes (CONTINUE)**

1a. Date of most recent scan (MM/DD/YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 1a: Enter the month, day, and year of the scan in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

1b. Are results of quantitative image analysis available?	☐ 0 No **(SKIP TO QUESTION 1d)** ☐ 1 Yes **(CONTINUE)** ☐ 9 Unknown **(SKIP TO QUESTION 1d)**

1c. Was there an MRI finding of hippocampal atrophy, according to your Center's standards for positivity? (REPORT MOST RECENT)	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 1c: Please use your Center's local standards for positivity to determine whether the participant had positive imaging findings for each of the Questions 1c, 2c, 2d, 2e, 5c. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0=No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the scan was repeated over time and the repeated scans were more than a month apart, report the result (+ or -) from the most recent scan. If the same scan was repeated multiple times at the same time, these are the most recent results available, and the results from these scans are conflicting, select 9=Unknown.

1d. Is an MRI available for data sharing?	☐ 0 No or unknown **(SKIP TO QUESTION 2)** ☐ 1 Yes **(COMPLETE QUESTIONS 1e – 1h)**
Questions 1e – 1h refer to MOST RECENT SCAN AVAILABLE:	
1e. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
1f. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes; ADNI version: _____ ☐ 9 Unknown

Question 1f: For information on Alzheimer's Disease Neuroimaging Initiative (ADNI) protocol, see: http://www.adni-info.org/Scientists/ADNIStudyProcedures.html.

1g. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other (SPECIFY): _____ ☐ 9 Unknown
1h. Field strength:	☐ 1 1.5T ☐ 2 3T ☐ 3 7T ☐ 4 Other (SPECIFY): _____ ☐ 9 Unknown

FDG-PET

2. Has the participant had at least one FDG-PET scan, obtained as part of the current evaluation or a previous evaluation?	☐ 0 **No or unknown (SKIP TO QUESTION 3)** ☐ 1 **Yes (CONTINUE)**

2a. Date of most recent scan (MM/DD/YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 2a: Enter the month, day, and year of the scan in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

2b. Are results of quantitative image analysis available?	☐ 0 No **(SKIP TO QUESTION 2f)** ☐ 1 Yes **(CONTINUE)** ☐ 9 Unknown **(SKIP TO QUESTION 2f)**

Questions 2c – 2e refer to MOST RECENT SCAN:

2c. Was there an FDG-PET finding of occipital hypometabolism consistent with LBD, according to your Center's standards for positivity?	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 2c: Please use your Center's local standards for positivity to determine whether the participant had positive imaging findings for each of the Questions 1c, 2c, 2d, 2e, 5c. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0=No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the scan was repeated over time and the repeated scans were more than a month apart, report the result (+ or -) from the most recent scan. If the same scan was repeated multiple times at the same time, these are the most recent results available, and the results from these scans are conflicting, select 9=Unknown.

2d. Was there an FDG-PET finding of temporoparietal hypometabolism suggestive of AD, according to your Center's standards for positivity?	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 2d: Please use your Center's local standards for positivity to determine whether the participant had positive imaging findings for each of the Questions 1c, 2c, 2d, 2e, 5c. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0=No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the scan was repeated over time and the repeated scans were more than a month apart, report the result (+ or -) from the most recent scan. If the same scan was repeated multiple times at the same time, these are the most recent results available, and the results from these scans are conflicting, select 9=Unknown.

2e. Was there an FDG-PET finding of cingulate island sign consistent with LBD, according to your Center's standards for positivity?	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 2e: Please use your Center's local standards for positivity to determine whether the participant had positive imaging findings for each of the Questions 1c, 2c, 2d, 2e, 5c. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0=No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the scan was repeated over time and the repeated scans were more than a month apart, report the result (+ or -) from the most recent scan. If the same scan was repeated multiple times at the same time, these are the most recent results available, and the results from these scans are conflicting, select 9=Unknown.

2f. Is an FDG-PET available for data sharing?	☐ 0 No or unknown **(SKIP TO QUESTION 3)** ☐ 1 Yes **(COMPLETE 2g – 2i)**
Questions 2g – 2i refer to MOST RECENT SCAN AVAILABLE:	
2g. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown

2h. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes; ADNI version: _____ ☐ 9 Unknown

Question 2h: For information on Alzheimer's Disease Neuroimaging Initiative (ADNI) protocol, see: http://www.adni-info.org/Scientists/ADNIStudyProcedures.html.

2i. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other (SPECIFY): _____ ☐ 9 Unknown
AMYLOID PET	
3. Has the participant had at least one amyloid PET scan, obtained as part of the current evaluation or a previous evaluation?	☐ 0 **No or unknown (SKIP TO QUESTION 4)** ☐ 1 **Yes (CONTINUE)**

3a. Date of most recent scan (MM/DD/YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 3a: Enter the month, day, and year of the scan in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

3b. Are results of quantitative image analysis available?	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
3c. Is an amyloid PET available for data sharing?	☐ 0 No **(SKIP TO QUESTION 4)** ☐ 1 Yes **(COMPLETE QUESTIONS 3d – 3g)** ☐ 9 Unknown **(SKIP TO QUESTION 4)**
Questions 3d – 3g refer to MOST RECENT SCAN AVAILABLE:	
3d. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
3e. Ligand used:	☐ 1 11C-PIB ☐ 2 18F-AV45 ☐ 3 Flutametamol ☐ 4 Other (specify): _____ ☐ 9 Unknown
3f. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes; ADNI version: _____ ☐ 9 Unknown

Question 3f: For information on Alzheimer's Disease Neuroimaging Initiative (ADNI) protocol, see: http://www.adni-info.org/Scientists/ADNIStudyProcedures.html.

3g. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other (SPECIFY): _____ ☐ 9 Unknown

TAU PET

4. Has the participant had at least one Tau PET scan, obtained as part of the current evaluation or a previous evaluation?	☐ 0 **No or unknown (SKIP TO QUESTION 5)** ☐ 1 **Yes (CONTINUE)**

4a. Date of most recent scan (MM/DD/YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 4a: Enter the month, day, and year of the scan in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

4b. Are results of quantitative image analysis available?	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
4c. Is a Tau PET available for data sharing?	☐ 0 No **(SKIP TO QUESTION 5)** ☐ 1 Yes **(COMPLETE QUESTIONS 4d – 4g)** ☐ 9 Unknown **(SKIP TO QUESTION 5)**
4d. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
4e. Ligand used:	☐ 1 18F-AV1451 (T807) ☐ 2 18F-THK5351 ☐ 3 Other, specify: _____

4f. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes; ADNI version: _____ ☐ 9 Unknown

Question 4f: For information on Alzheimer's Disease Neuroimaging Initiative (ADNI) protocol, see: http://www.adni-info.org/Scientists/ADNIStudyProcedures.html.

4g. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other (SPECIFY): _____ ☐ 9 Unknown

DaTScan

5. Has the participant had at least one DaTScan scan, obtained as part of the current evaluation or a previous evaluation?	☐ 0 **No or unknown (END FORM HERE)** ☐ 1 **Yes (CONTINUE)**

5a. Date of scan (MM/DD/YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 5a: Enter the month, day, and year of the scan in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

5b. Are results of quantitative image analysis available?	☐ 0 No **(END FORM HERE)** ☐ 1 Yes **(CONTINUE)** ☐ 9 Unknown **(END FORM HERE)**

5c. Were there abnormal DaTScan findings consistent with LBD, according to your Center's standards for positivity? (REPORT MOST RECENT)	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 5c: Please use your Center's local standards for positivity to determine whether the participant had positive imaging findings for each of the Questions 1c, 2c, 2d, 2e, 5c. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0=No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the scan was repeated over time and the repeated scans were more than a month apart, report the result (+ or -) from the most recent scan. If the same scan was repeated multiple times at the same time, these are the most recent results available, and the results from these scans are conflicting, select 9=Unknown.

Form E3L: Other Labs and Findings

Polysomnography

1. Has the participant had at least one polysomnography, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 2)** ☐ 1 Yes

1a. Date of most recent polysomnography (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 1a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

1b. Was there polysomnographic confirmation of REM sleep without atonia, +/– dream enactment behavior, consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 1b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

1c. Is a polysomnography available for data sharing?	☐ 0 No or unknown ☐ 1 Yes

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

Cardiac-MIBG scintigraphy

2. Has the participant had at least one cardiac-MIBG scintigraphy, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 3)** ☐ 1 Yes

2a. Date of most recent cardiac-MIBG scintigraphy (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 2a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

2b. Were there abnormal (low uptake) MIBG myocardial scintigraphy results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 2b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

2c. Is a cardiac-MIBG available for data sharing?	☐ 0 No or unknown ☐ 1 Yes, raw data available ☐ 2 Yes, processed data available ☐ 3 Yes, both raw and processed data available

Anosmia test (e.g., UPSIT)

3. Has the participant had at least one anosmia test, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 4)** ☐ 1 Yes

3a. Date of most recent anosmia test (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	___ ___ / ___ ___ / ___ ___ ___ ___

Question 3a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

3b. Were the anosmia test results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 3b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

3c. Are anosmia test data available for sharing?	☐ 0 No or unknown **(SKIP TO QUESTION 4)** ☐ 1 Yes
3d. Which test was done (that is available for sharing)?	☐ 1 University of Pennsylvania Smell Identification Test (UPSIT) ☐ 2 Brief-smell identification test (B-SIT) ☐ 3 Sniffin Sticks ☐ 4 Other (SPECIFY): _____

Electroencephalogram (EEG)

4. Has the participant had at least one electroencephalogram, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 5)** ☐ 1 Yes
4a. Date of most recent electroencephalogram (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 4a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

4b. Was there prominent posterior slow wave activity on EEG with periodic fluctuations in the prealpha/theta range, consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 4b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

4c. Is an electroencephalogram available for data sharing?	☐ 0 No or unknown ☐ 1 Yes, raw data available ☐ 2 Yes, processed data available ☐ 3 Yes, both raw and processed data available

Multiple sleep latency test (MSLT)

5. Has the participant had at least one MSLT, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 6)** ☐ 1 Yes

5a. Date of most recent MSLT (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 5a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

5b. Were the MSLT results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 5b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

5c. Are MSLT data available for sharing?	☐ 0 No or unknown ☐ 1 Yes

Tilt table test

6. Has the participant had at least one tilt table test, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 7)** ☐ 1 Yes
6a. Date of most recent tilt table test (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 6a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

6b. Were the tilt table test results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 6b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

6c. Are tilt table test data available for sharing?	☐ 0 No or unknown ☐ 1 Yes

Quantitative sudomotor axon reflex test (QSART)

7. Has the participant had at least one QSART, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 8** ☐ 1 Yes
7a. Date of most recent QSART (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 7a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

7b. Were the QSART results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 7b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

7c. Are QSART data available for sharing?	☐ 0 No or unknown ☐ 1 Yes

Thermoregulatory sweat test

8. Has the participant had at least one thermoregulatory sweat test, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(SKIP TO QUESTION 9)** ☐ 1 Yes

8a. Date of most recent thermoregulatory sweat test (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 8a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

8b. Were the thermoregulatory sweat test results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 8b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

8c. Are thermoregulatory sweat test data available for sharing?	☐ 0 No or unknown ☐ 1 Yes

Computerized gait testing

9. Has the participant had at least one computerized gait testing, obtained as part of the current evaluation or a previous evaluation?	☐ 0 No or unknown **(END FORM HERE)** ☐ 1 Yes

9a. Date of most recent computerized gait testing (MM/DD/YYYY): *NOTE: A value of 99=Unknown is permissible for day only.*	__ __ / __ __ / __ __ __ __

Question 9a: Enter the month, day, and year of the assay/test in the specified numerical format (e.g. March 1, 2010 would be entered as 03/01/2010. If the exact day is unknown, enter 99 in the appropriate field (e.g. March 2010 would be 03/99/2010).

9b. Were the computerized gait testing results consistent with LBD, according to your Center's standards for positivity? **(REPORT MOST RECENT)**	☐ 0 No ☐ 1 Yes ☐ 8 Results not available ☐ 9 Unknown

Question 9b: Please use your Center's local standards for positivity to determine whether the participant had positive laboratory findings for each of the Questions 1b, 2b, 3b, 4b, 5b, 6b, 7b, 8b, 9b. If the results were positive according to your local standards, select 1=Yes. If the results were negative, select 0= No. If the results were ambiguous according to your Center's standard cutoff values (i.e., "too close to call"), select 0=No. If the results are not available at this time or not available for sharing, select 8=Results not available.

If the specific laboratory assay or test was repeated over time and the repeated assays/tests were more than a month apart, report the result (+ or -) from the most recent assay/test. If the same assay/test was repeated multiple times at the same time, these are the most recent results available, and the results from these assays/tests are conflicting, select 9=Unknown.

9c. Are computerized gait testing data available for sharing?	☐ 0 No or unknown ☐ 1 Yes, raw data available ☐ 2 Yes, processed data available ☐ 3 Yes, both raw and processed data available

Form D1L: Clinical DLB and PD Features

DETERMINING PRESENCE OR ABSENCE FOR FORM D1L:

Select **0=Absent** if a symptom is not reported by the participant/co-participant as present during the last 6 months, and/or is determined absent by the clinician during the last 6 months based on clinician judgment after testing/use of an instrument, medical records, and/or examination.

Select **2=Definitely Present** if the clinician judges the symptom to be present on at least 3 occasions during the last 6 months based on participant/co-participant report, and/or if there is confirmation through testing/use of an instrument, medical records, examination, and/or existing or new diagnosis in the past 6 months.

Select **1=Questionably Present** if after reviewing all available information, the clinician finds that the evidence is mixed; the findings from interview, testing/use of an instrument, medical records, and/or examination were not certain enough to indicate as Definitely Present or Absent; or if the only evidence is participant/co-participant report that the symptom was present less than 3 times over the past 6 months and based on clinical judgment.

Select **8=Not evaluated** if the symptom was not assessed at this visit.

Gateway question for cognitive symptoms

1. Is an acquired disorder of cognition a prominent element of the clinical presentation of the participant? (I.e., at least one of the characteristics described in Questions 1a–1e is "Definitely present.")

☐ 0 No **(SKIP TO QUESTION 2)**

☐ 1 Yes

An acquired disorder of cognition is definitely present if through cognitive testing and/or clinician judgment it is determined that the participant is experiencing significant changes in his/her usual or customary cognitive functioning — i.e., is experiencing deficits in episodic memory, language, attention, executive function, and/or visuoperceptual ability.

Characterizing cognitive symptoms

Please indicate whether any of the features listed below are present during the current examination.	Absent	Questionably present	Definitely present	Not evaluated
1a. Episodic memory deficits	☐ 0	☐ 1	☐ 2	☐ 8
1b. Language deficits	☐ 0	☐ 1	☐ 2	☐ 8
1c. Attention deficits	☐ 0	☐ 1	☐ 2	☐ 8
1d. Executive deficits	☐ 0	☐ 1	☐ 2	☐ 8
1e. Visuoperceptual deficits	☐ 0	☐ 1	☐ 2	☐ 8

Gateway question for motor symptoms

2. Is an acquired disorder of movement a prominent element of the clinical presentation of the participant? (I.e., at least one of the characteristics described in Questions 2a–2h is "Definitely present.")

☐ 0 No **(SKIP TO QUESTION 3)**

☐ 1 Yes

An acquired disorder of movement is definitely present if through a physical and/or motor exam it is determined that the participant is experiencing significant changes in his/her usual or customary motor function — i.e., is experiencing at least one of the following: bradykinesia, rigidity, rest tremor, postural tremor, action tremor, myoclonus, gait abnormality, and/or postural instability.

Characterizing motor symptoms

Please indicate whether any of the features listed below are present during the current examination.	Absent	Questionably present	Definitely present	Not evaluated
2a. Bradykinesia	☐ 0	☐ 1	☐ 2	☐ 8
Bradykinesia includes combining slowness, hesitancy, decreased arm swing, small amplitude, and poverty of movement in general.				
2b. Rigidity (with or without cogwheel characteristics)	☐ 0	☐ 1	☐ 2	☐ 8
Rigidity with or without cogwheel characteristics is judged based on passive movement of major joints with patient relaxed in sitting position.				
2c. Rest tremor	☐ 0	☐ 1	☐ 2	☐ 8
Rest tremor is observed in a completely relaxed part of the body.				
2d. Postural tremor	☐ 0	☐ 1	☐ 2	☐ 8
Postural tremor is observed when a part of the body is maintained against gravity.				
2e. Action tremor	☐ 0	☐ 1	☐ 2	☐ 8
Action tremor is observed at movement.				
2f. Myoclonus	☐ 0	☐ 1	☐ 2	☐ 8
Myoclonus is a sudden shock-like twitching of muscles or parts of muscles without any rhythm or pattern.				
2g. Gait abnormality	☐ 0	☐ 1	☐ 2	☐ 8
Gait abnormality includes slowing of gait, shuffling, festination, unilateral or bilateral decreased arm swing and/or tremor, slowness and difficulty on turning, and/or freezing during walking.				
2h. Postural instability	☐ 0	☐ 1	☐ 2	☐ 8
Postural instability involves inadequate response to sudden, strong posterior displacement produced by pull on shoulders while patient is erect with eyes open and feet slightly apart (patient is prepared).				

Gateway question for behavioral symptoms

3. Is an acquired disorder of behavior a prominent element of the clinical presentation of the participant? (I.e., at least one of the characteristics described in Questions 3a–3e is "Definitely present.")

☐ 0 No **(SKIP TO QUESTION 4)**

☐ 1 Yes

An acquired disorder of behavior is definitely present if, through observation, previous clinical diagnosis, and/or clinician judgment based on participant and co-participant report, it is determined that the participant is experiencing significant changes in his/her usual or customary behavior — i.e., is experiencing at least one of the following: depression, apathy, anxiety, hallucinations, or delusions.

Characterizing behavioral symptoms

Please indicate whether any of the features listed below are present during the current examination.	Absent	Questionably present	Definitely present	Not evaluated
3a. Depression	☐ 0	☐ 1	☐ 2	☐ 8
3b. Apathy	☐ 0	☐ 1	☐ 2	☐ 8
3c. Anxiety	☐ 0	☐ 1	☐ 2	☐ 8
3d. Hallucinations	☐ 0	☐ 1	☐ 2	☐ 8
3e. Delusions	☐ 0	☐ 1	☐ 2	☐ 8

Gateway question for autonomic or constitutional symptoms

4. Is an acquired disorder of autonomic or constitutional features a prominent element of the clinical presentation of the participant? (I.e., at least one of the characteristics described in Questions 4a–4l is "Definitely present.")

☐ 0 No **(SKIP TO QUESTION 5)**

☐ 1 Yes

Characterizing autonomic or constitutional symptoms

Please indicate whether any of the features listed below are present during the current examination.	Absent	Questionably present	Definitely present	Not evaluated
4a. REM sleep behavior disorder	☐ 0	☐ 1	☐ 2	☐ 8
4b. Obstructive sleep apnea	☐ 0	☐ 1	☐ 2	☐ 8
4c. Periodic leg movements of sleep	☐ 0	☐ 1	☐ 2	☐ 8
4d. Restless leg syndrome	☐ 0	☐ 1	☐ 2	☐ 8
4e. Excessive daytime sleepiness	☐ 0	☐ 1	☐ 2	☐ 8
4f. Cognitive fluctuations	☐ 0	☐ 1	☐ 2	☐ 8
4g. Orthostatic hypotension	☐ 0	☐ 1	☐ 2	☐ 8

Orthostatic hypotension is a change in blood pressure and heart rate that is consistent with orthostatic hypotension — i.e., drop of 20mmHg drop in systolic blood pressure (SBP) or 10mmHg drop in diastolic blood pressure (DBP) at least 1 minute after going from supine to sitting or supine to standing.

4h. Constipation	☐ 0	☐ 1	☐ 2	☐ 8
4i. Hyposmia	☐ 0	☐ 1	☐ 2	☐ 8
4j. Falls	☐ 0	☐ 1	☐ 2	☐ 8
4k. Syncope	☐ 0	☐ 1	☐ 2	☐ 8
4l. Severe sensitivity to anti-psychotic agents	☐ 0	☐ 1	☐ 2	☐ 8

Severe sensitivity to anti-psychotic agents (neuroleptics) is characterized by the presence of at least one of the typical signs following drug use: worsening cognition, heavy sedation, increased or possibly irreversible parkinsonism, or symptoms consistent with neuroleptic malignant syndrome (NMS) (e.g., severe fever and muscle rigidity).

Cognitive status and etiology

5. What is the participant's cognitive status?

☐ 1 Normal cognition
☐ 2 Cognitively impaired, not MCI
☐ 3 MCI
☐ 4 Dementia

The participant's cognitive status reported in Question 5 should be consistent with what is reported on UDS Form D1 (Clinical Diagnosis).

Normal cognition is diagnosed if the participant:

- Has global CDR=0 and/or neuropsychological testing within the normal range
- Has normal behavior (does not exhibit behavior sufficient to diagnosis MCI or dementia due to FTLD or LBD)

Dementia is diagnosed if the participant 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.*

MCI is diagnosed if the participant:

- Has a cognitive complaint
- Has cognition that is not normal for age
- Is not demented
- Has experienced cognitive decline
- Has essentially normal functional activities

"Cognitively impaired, not MCI" is diagnosed if the participant:

- Is cognitively impaired
- Has a presentation, test results, symptoms, and clinical evaluation that are not consistent with MCI (and not consistent with normal cognition or dementia)

6. Which etiologic diagnosis best characterizes the participant?

- ☐ 1 Dementia with Lewy bodies
- ☐ 2 Parkinson's disease
- ☐ 3 Alzheimer's disease
- ☐ 4 Vascular disease
- ☐ 5 FTLD
- ☐ 6 Other
- ☐ 8 Not applicable — no neurodegenerative disease and no cognitive impairment

Please refer to the diagnostic criteria below for DLB, PD, AD, CVD, and FTLD as listed below to determine the primary etiology.

DLB CRITERIA

Revised (2017[1]) criteria for the clinical diagnosis of dementia with Lewy bodies (DLB):

1. Essential for a diagnosis of DLB is dementia, defined as a progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational functions, or with usual daily activities. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression. Deficits on tests of attention, executive function, and visuoperceptual ability may be especially prominent and occur early.
2. Core clinical features (the first 3 typically occur early and may persist throughout the course):
   - Fluctuating cognition with pronounced variations in attention and alertness.
   - Recurrent visual hallucinations that are typically well-formed and detailed.
   - REM sleep behavior disorder, which may precede cognitive decline.
   - One or more spontaneous cardinal features of parkinsonism: these are bradykinesia (defined as slowness of movement and decrement in amplitude or speed), rest tremor, or rigidity.
3. Supportive clinical features:
   - Severe sensitivity to antipsychotic agents; postural instability.
   - Repeated falls.
   - Syncope or other transient episodes of unresponsiveness.
   - Severe autonomic dysfunction, e.g., constipation, orthostatic hypotension, urinary incontinence.
   - Hypersomnia.
   - Hyposmia.
   - Hallucinations in other modalities.
   - Systematized delusions.
   - Apathy.
   - Anxiety.
   - Depression.
4. Indicative biomarkers:
   - Reduced dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET.
   - Abnormal (low-uptake) 123iodine-MIBG myocardial scintigraphy.
   - Polysomnographic confirmation of REM sleep without atonia.
5. Supportive biomarkers:
   - Relative preservation of medial temporal lobe structures on CT/MRI scan.
   - Generalized low uptake on SPECT/PET perfusion/metabolism scan with reduced occipital activity 6 the cingulate island sign on FDG-PET imaging.
   - Prominent posterior slow-wave activity on EEG with periodic fluctuations in the pre-alpha/theta range.

6. Probable DLB can be diagnosed if:
   a. Two or more core clinical features of DLB are present, with or without the presence of indicative biomarkers, or
   b. Only one core clinical feature is present, but with one or more indicative biomarkers. Probable DLB should not be diagnosed on the basis of biomarkers alone.

7. Possible DLB can be diagnosed if:
   a. Only one core clinical feature of DLB is present, with no indicative biomarker evidence, or
   b. One or more indicative biomarkers is present but there are no core clinical features.

8. DLB is less likely:
   - In the presence of any other physical illness or brain disorder including cerebrovascular disease, sufficient to account in part or in total for the clinical picture, although these do not exclude a DLB diagnosis and may serve to indicate mixed or multiple pathologies contributing to the clinical presentation.
   - If parkinsonian features are the only core clinical feature and appear for the first time at a stage of severe dementia.

DLB should be diagnosed when dementia occurs before or concurrently with parkinsonism. The term Parkinson disease dementia (PDD) should be used to describe dementia that occurs in the context of well-established Parkinson disease. In a practice setting the term that is most appropriate to the clinical situation should be used, and generic terms such as Lewy body disease are often helpful. In research studies in which distinction needs to be made between DLB and PDD, the existing 1-year rule between the onset of dementia and parkinsonism continues to be recommended.

(For more information on the criteria above, visit the website of the Lewy Body Dementia Association at https://www.lbda.org/newdlbcriteria.)

[1] Guidebook updated September 19, 2017 to reflect the recommendations for the clinical diagnosis of DLB by the Dementia with Lewy Bodies Consortium.

McKeith IF, Boeve BF, Dickson DW, et al. Diagnosis and management of dementia with Lewy bodies: Fourth consensus report of the DLB Consortium, Neurology 2017; 89: 88-100.

PD CRITERIA

Use the following criteria, excerpted from SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders (Litvan et al., 2003):

UK PARKINSON'S DISEASE SOCIETY BRAIN BANK CLINICAL DIAGNOSTIC CRITERIA

Inclusion criteria	Exclusion criteria	Supportive criteria
Bradykinesia (slowness of initiation of voluntary movement with progressive reduction in speed and amplitude of repetitive actions); And at least one of the following: • Muscular rigidity. • 4- to 6-Hz rest tremor. • Postural instability not caused by primary visual, vertibular, cerebellar, or proprioceptive dysfunction.	History of repeated strokes with stepwise progression of parkinsonian features. History of repeated head injury. History of definite encephalitis. Oculogyric crises. Neuroleptic treatment at onset of symptoms. More than one affected relative. Sustained remission. Strictly unilateral features after 3 years. Supranuclear gaze palsy. Cerebellar signs. Early severe autonomic involvement. Early severe dementia with disturbances of memory, language, and praxis. Babinski sign. Presence of cerebral tumor or communicating hydrocephalus on CT scan. Negative response to large doses of levodopa (if malabsorption excluded). MPTP exposure.	(Three or more required for diagnosis of definite PD): • Unilateral onset. • Rest tremor present. • Progressive disorder. • Persistent asymmetry affecting side of onset most. • Excellent response (70%–100%) to levodopa. • Severe levodopa-induced chorea. • Levodopa response for 5 years or more. • Clinical course of 10 years or more.

UK = United Kingdom; PD = Parkinson's disease; CT = computed tomography.

AD CRITERIA

The AD dementia criteria listed below are excerpted and condensed from the 2011 NIA-AA criteria for AD dementia (McKhann et al., 2011). The diagnosis of dementia due to Alzheimer's disease: Recommendations from the National Institute on Aging and the Alzheimer's Association workgroups. See the original paper for details.

A. Probable AD dementia is diagnosed when the patient:

1. Meets criteria for dementia, and has the following characteristics:
2. Insidious onset. Symptoms have a gradual onset over months to years; and
3. Clear-cut history of worsening of cognition by report or observation; and
4. The initial and most prominent cognitive deficits are evident on history and examination in one of the following categories.
   (1) Amnestic disorder: The most common syndromic presentation of AD dementia.
   (2) Non-amnestic disorders:
   - Language disorder
   - Visuospatial disorder
   - Executive and behavioral disorder
5. Exclusions: The diagnosis of probable AD dementia should not be applied when there is evidence of:
   (a) substantial concomitant cerebrovascular disease or
   (b) core features of dementia with Lewy bodies other than dementia itself; or
   (c) prominent features of behavioral variant frontotemporal dementia; or

(d) prominent features of semantic variant primary progressive aphasia or non-fluent/agrammatic variant primary progressive aphasia; or
(e) evidence for another concurrent, active neurological disease, or a non-neurological medical co-morbidity or medication use that could have a substantial impact on cognition.

B. Possible AD dementia is diagnosed when the patient meets one of the two following criteria:

1. Atypical course: Meets the core clinical criteria (1) and (4) (above) for probable AD dementia, but either had a sudden onset of cognitive impairment or demonstrates insufficient historical detail or objective cognitive documentation of progressive decline, or
2. Etiologically mixed presentation: Meets all core clinical criteria (1) through (4) (above) for probable AD dementia but has evidence of:
   (a) concomitant cerebrovascular disease or
   (b) features of dementia with Lewy bodies other than the dementia itself; or
   (c) evidence for another neurological disease or a non-neurological medical co-morbidity or medication use that could have a substantial impact on cognition.

McKhann GM, Knopman DS, Chertkow H, et al. The diagnosis of dementia due to Alzheimer's disease: recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. Alzheimers Dement. 2011;7(3):263-269.

The following table is excerpted from the 2011 NIA-AA criteria for MCI due to dementia (Albert et al., 2011):

Summary of clinical and cognitive evaluation for MCI due to AD

Establish clinical and cognitive criteria
Cognitive concern reflecting a change in cognition reported by patient or informant or clinician (i.e., historical or observed evidence of decline over time)
Objective evidence of impairment in one or more cognitive domains, typically including memory (i.e., formal or bedside testing to establish level of cognitive function in multiple domains)
Largely preserved independence in functional abilities
Not demented
Examine etiology of MCI consistent with AD pathophysiological process
Rule out vascular, traumatic, medical causes of cognitive decline, where possible
Provide evidence of longitudinal decline in cognition, when feasible
Report history consistent with AD genetic factors, where relevant

Abbreviations: AD, Alzheimer's disease; MCI, mild cognitive impairment

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"Alzheimer's and Dementia, 7/3, Albert MS, DeKosky ST, Dickson D, Dubois B, Feldman HH, Fox NC, Gamst A, Holtzman DM, Jagust WJ, Petersen RC, Snyder PJ, Carrillo MC, Thies B, Phelps CH, The diagnosis of mild cognitive impairment due to Alzheimer's disease: recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease, 270-279, 2011, with permission from Elsevier http://www.sciencedirect.com/science/article/pii/S155252601100104X."

CVD CRITERIA

Significant vascular brain injury includes either:

- CLINICAL EVIDENCE of symptomatic stroke (i.e., abrupt onset of focal neurological signs)

– OR –

- NEUROIMAGING EVIDENCE of one or more of the following:
  – cystic infarcts (large or small)
  – significant white matter changes (Grade 7–8+ on Cardiovascular Health Study Scale)

– intraparenchymal hemorrhage
– multiple microbleeds

If the subject has no clinical evidence of symptomatic stroke and neuroimaging studies do not indicate evidence of significant vascular brain injury.

FTLD CRITERIA (PPA, BVFTD, OTHER)

ROOT DIAGNOSIS OF PRIMARY PROGRESSIVE APHASIA (PPA)[1]
All three core criteria must be present:

1. Most prominent clinical feature is a new and progressive difficulty with language (i.e., retrieving, using, repeating, sequencing, or understanding words).
2. The language disorder (aphasia) should be the most prominent deficit at symptom onset and for the initial phases of the disease.
3. All causes other than neurodegeneration are excluded.

[1]Mesulam, M.-M., 2003. Primary progressive aphasia: A language-based dementia. New England Journal of Medicine 348, 1535-1542.

International consensus criteria for behavioural variant FTD (FTDC)

I. Neurodegenerative disease

The following symptom must be present to meet criteria for bvFTD.

A. Shows progressive deterioration of behaviour and/or cognition by observation or history (as provided by a knowledgeable informant).

II. Possible bvFTD

Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.

A Early* behavioural disinhibition [one of the following symptoms (A1–A3) must be present]:
- A1. Socially inappropriate behaviour
- A2. Loss of manners or decorum
- A3. Impulsive, rash or careless actions

B. Early apathy or inertia [one of the following symptoms (B1–B2) must be present]:
- B1. Apathy
- B2. Inertia

C. Early loss of sympathy or empathy [one of the following symptoms (C1–C2) must be present]:
- C1. Diminished response to other people's needs and feelings
- C2. Diminished social interest, interrelatedness or personal warmth

D Early perseverative, stereotyped or compulsive/ritualistic behaviour [one of the following symptoms (D1–D3) must be present]:
- D1. Simple repetitive movements
- D2. Complex, compulsive or ritualistic behaviours
- D3. Stereotypy of speech

E. Hyperorality and dietary changes [one of the following symptoms (E1–E3) must be present]:
- E1. Altered food preferences
- E2. Binge eating, increased consumption of alcohol or cigarettes
- E3. Oral exploration or consumption of inedible objects

F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions [all of the following symptoms (F1–F3) must be present]:
- F1. Deficits in executive tasks
- F2. Relative sparing of episodic memory
- F3. Relative sparing of visuospatial skills

III. Probable bvFTD

All of the following symptoms (A–C) must be present to meet criteria.

A. Meets criteria for possible bvFTD
B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
C. Imaging results consistent with bvFTD [one of the following (C1–C2) must be present]:
   C1. Frontal and/or anterior temporal atrophy on MRI or CT
   C2. Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT

IV. Behavioural variant FTD with definite FTLD pathology

Criterion A and either criterion B or criterion C must be present to meet criteria.

A. Meets criteria for possible or probable bvFTD
B. Histopathological evidence of FTLD on biopsy or at post-mortem
C. Presence of a known pathogenic mutation

V. Exclusionary criteria for bvFTD

Criteria A and B must be answered negatively for any bvFTD diagnosis. Criterion C can be positive for possible bvFTD but must be negative for probable bvFTD.

A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
B. Behavioural disturbance is better accounted for by a psychiatric diagnosis
C. Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process

**As a general guideline, "early" refers to symptom presentation within the first 3 years.*
bvFTD = behavioral variant FTD

PSP CRITERIA

Use the following criteria to diagnose PSP (adapted from Bensimon et al., 2009)

Inclusion criteria

ALL OF THE FOLLOWING:

- Age at disease onset ≥30 years;
- Akinetic-rigid syndrome;
- Postural instability or falls (within 3 years from disease onset);
- Supranuclear ophthalmoplegia.

Exclusion criteria

ANY OF THE FOLLOWING:

- Cerebellar ataxia;
- Evidence of any other neurological disease that could explain signs;
- History of repeated strokes with stepwise progression of parkinsonian features;
- Idiopathic Parkinson's disease;
- Oculogyric crises;
- Significant other neurological disease on CT-scan/MRI;
- Signs of corticobasal degeneration;
- Signs of lewy body disease;
- Symptomatic autonomic dysfunction;
- Tremor at rest.

CBD CRITERIA

Proposed clinical phenotypes (syndromes) associated with the pathology of corticobasal degeneration (CBD)

Syndrome	Features
Probable corticobasal syndrome	**Asymmetric presentation of TWO OF:** a) limb rigidity or akinesia, b) limb dystonia, c) limb myoclonus; **PLUS TWO OF:** d) orobuccal or limb apraxia, e) cortical sensory deficit, f) alien limb phenomena (more than simple levitation).
Possible corticobasal syndrome	**May be symmetric; ONE OF:** a) limb rigidity or akinesia, b) limb dystonia, c) limb myoclonus; **PLUS ONE OF:** d) orobuccal or limb apraxia, e) cortical sensory deficit, f) alien limb phenomena (more than simple levitation).

[1] Armstrong, MJ, Litvan I, et al. *Criteria for the diagnosis of corticobasal degeneration*. Neurology 2013;80;496.

ALS CRITERIA

Use the following criteria, adapted from El Escorial revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis (Brooks et al., 2000)[1]:

Requirements for the diagnosis of amyotrophic lateral sclerosis

The diagnosis of ALS requires the PRESENCE of:

- Evidence of lower motor neuron (LMN) degeneration by clinical, electrophysiological or neuropathologic examination;
- Evidence of upper motor neuron (UMN) degeneration by clinical examination; **and**
- Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination, **together with** B1 and B2 in next column.

The diagnosis of ALS requires the ABSENCE of:

- Electrophysiological or pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration; **and**
- Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs.

[1] Brooks BR, Miller RG, Swash M, Munsat TL, Diseases WFoNRGoMN. El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. Amyotroph Lateral Scler Other Motor Neuron Disord. 2000;1(5):293-299.