



NACC UNIFORM DATA SET **FTLD MODULE**
Coding Guidebook for FVP

Version 3.0, March 2015

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ADC Program (David Knopman, MD, Chair) and the
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This publication was funded by the National Institutes
of Health through the National Institute on Aging
(Cooperative Agreement U01 AG016976).

Revisions since UDS3 implementation (March 15, 2015)

| Date yyyy-mm-dd | Description | Form(s) affected | Question(s) affected | Data element(s) affected |
|--------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------|----------------------|-----------------------------|
| 2016-10-26 | Instructions for Form B9F changed. Original text: "The neuropsychological test batteries and imaging results (from both the FTL D Module and the Uniform Data Set) should not be used to determine responses for this form, but should be used to make the official clinical diagnosis on Form D1 of the UDS. Conclusions should be based on information obtained through subject, co-participant, medical records, and/or observation." New text: "Conclusions should be based on information obtained through subject, co-participant, medical records, observation, imaging, and/or neuropsychological testing." | B9F | All | N/A |
| 2015-10-22 | Coding corrected for PARENT (=4) and SIBLING (=5) under "Relationship to subject" | C5F | N/A | FTDINFRE |

FTLD Module to the Uniform Data Set Coding Guidebook for the Follow-up Visit Packet

INTRODUCTION

The FTLD Module to the UDS is designed for:

- persons with primary diagnoses of primary progressive aphasia (PPA) and behavioral variant frontotemporal degeneration (bvFTD)
- persons with mild cognitive impairment who are thought to be at risk for developing PPA or bvFTD
- persons with other neurodegenerative diseases such as PSP, CBS, ALS that have features that overlap with bvFTD or PPA
- control subjects, at the Center's discretion

In addition, the Module may be useful in cognitively normal persons and in persons whose primary diagnoses are Alzheimer's disease, vascular dementia, or dementia with Lewy bodies and who have prominent linguistic or behavioral problems.

IMPORTANT NOTES

- **Timing** — The FTLD Module evaluation is intended to be completed as part of a UDS visit. If the UDS evaluation and the FTLD evaluation are separated into two days, please complete the FTLD 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 FTLD) from both days.
- **IVP vs. FVP** — When a UDS enrollee is being given the FTLD Module evaluation for the first time, you should use the FTLD Module Initial Visit Packet, even if you are using the UDS Follow-up Visit Packet.

How to read the Guidebook

The Guidebook features a reproduction of each form in the FTLD Module, interspersed with explanatory notes and references. Throughout this document, all explanatory and reference text are on white background. To make it easier to locate the Guidebook material, the reproductions of the forms themselves have been given a blue background.

Guide to abbreviations

| | |
|-------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| AD | Alzheimer's disease |
| ADC | Alzheimer's Disease Center, any of 30 Centers across the United States participating in the Alzheimer's Disease Centers Program conducted by NIA |
| ADNI | Alzheimer's Disease Neuroimaging Initiative |
| ALS | Amyotrophic lateral sclerosis |
| bvFTD | Behavioral variant frontotemporal dementia |
| CBD | Corticobasal degeneration |
| DLB | Dementia with Lewy bodies |
| FTLD | Frontotemporal lobar degeneration |
| FTLD Module | A collection of data concerning FTLD on subjects in the NACC Uniform Data Set and appended to the UDS |
| FVP | Follow-up Visit Packet, the set of forms completed at a subject's second and subsequent evaluations for submission to NACC |
| IVP | Initial Visit Packet, the set of forms completed at a subject's initial evaluation for submission to NACC |
| MCI | Mild cognitive impairment |
| MMSE | Mini-mental state examination |
| MND | Motor neuron disease |
| NACC | National Alzheimer's Coordinating Center, funded by NIA and charged with collecting data from the ADCs |
| NIA | National Institute on Aging, one of the U.S. National Institutes of Health |
| PLS | Primary lateral sclerosis |
| PPA | Primary progressive aphasia |
| SMA | Spinal muscular atrophy |
| UDS | Uniform Data Set, the longitudinal database maintained by NACC; the other components of the NACC database are the Minimum Data Set (MDS) and the Neuropathologic Data Set (NP) |
| UPDRS | Unified Parkinson's Disease Rating Scale |
| VaD | Vascular dementia |

A note on form numbering

Each NACC form has a unique two- to four-character number (e.g., B9, E2F, Z1, A3aF). For forms that are part of the FTLD Module to the UDS, the **last character is F** (with the exception of Form A3a). As in the UDS, the **first character** of the form number indicates what kind of information is collected by the form:

- A: Family history (genetic) data
- B: Clinical data
- C: Neuropsychologic data
- E: Imaging data
- Z: Used only for the Form Checklist

Form Z1F: Form Checklist

INSTRUCTIONS: This form is to be completed by clinic staff.

NACC expects and intends that all FTLD forms be attempted on all subjects being evaluated for the FTLD Module of the UDS, but we realize this may be impossible when the subject is terminally ill, or when there is no co-participant, or for other reasons. Nevertheless, NACC **requires** that Forms B3F, B9F, C1F, C2F, C3F, E2F, and E3F be submitted for a subject to be included in the FTLD Module of the UDS database, even though these forms may include some items with missing data.

For forms **not** designated as required, if it is not feasible to collect all or almost all of the data elements for a subject, and the ADC therefore decides not to attempt collection of those data, an explanation should be provided. Please document this decision by including the appropriate explanatory code and any additional comments.

KEY: If the specified form was not completed, please enter one of the following codes:

95=Physical problem 96=Cognitive/behavior problem 97=Other problem 98=Verbal refusal 99=Unknown or inadequate info

| | Description | Submitted: | | If not submitted, specify reason (see key, above) | Comments (provide if needed) |
|------------|---------------------------------------------|----------------------------|----------------------------|---------------------------------------------------------|---------------------------------|
| | | Yes | No | | |
| A3a | Record of Consent for Biologic Specimen Use | <input type="checkbox"/> 1 | <input type="checkbox"/> 0 | ___ ___ | |
| B3F | Supplemental UPDRS | Required | | | |
| B9F | Clinical PPA and bvFTD Features | Required | | | |
| C1F | Neuropsychological Battery Summary Scores | Required | | | |
| C2F | Social Norms Questionnaire | Required | | | |
| C3F | Social Behavior Observer Checklist | Required | | | |
| C4F | Behavioral Inhibition Scale | <input type="checkbox"/> 1 | <input type="checkbox"/> 0 | ___ ___ | |
| C5F | Interpersonal Reactivity Index | <input type="checkbox"/> 1 | <input type="checkbox"/> 0 | ___ ___ | |
| C6F | Revised Self-monitoring Scale | <input type="checkbox"/> 1 | <input type="checkbox"/> 0 | ___ ___ | |
| E2F | Imaging Available | Required | | | |
| E3F | Imaging in Diagnosis | Required | | | |

Select **1=Yes** if the specified form was completed for the subject during this visit. If a form is not designated as required and is not submitted, select **0=No**, enter the appropriate reason code from the Key, and provide a written explanation in the Comments field.

Form A3a: Record of Consent for Biologic Specimen Use

INSTRUCTIONS: This form is to be completed by clinic staff responsible for obtaining consents, based on an existing consent at clinic. For additional clarification, see FTLD Coding Guidebook for Follow-up Visit Packet, Form A3aF.

One of these forms will be completed for each relative who provides a specimen.

1. What relative's consent is being recorded on this form?

NOTE: "Unknown" (9999) is not a permissible value for sibling's or child's birth year. If birth year is unknown, please provide an approximate year on **UDS Initial Visit Form A3** so that the sibling or child ends up in correct birth order relative to the other siblings or children.

"Sibling's birth year" or "child's birth year" on this form MUST agree with the birth year listed for that person on UDS Initial Visit or UDS Follow-up Visit Form A3.

- 1 Mother
- 2 Father
- 3 Sibling (sibling's birth year: _____)
- 4 Child (child's birth year: _____)

Birth year for a given child or sibling should be constant across the following forms:

- Uniform Data Set (UDS) Initial Visit Form A3 or UDS Follow-up Visit Form A3 (if applicable)
- FTLD Module Initial Visit Form A3aF or FTLD Module Follow-up Visit Form A3aF (if applicable)

Please indicate that the above relative provided consent for the following. The wording need not be identical but should explicitly express the same points.

- | | | | |
|------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------|--------------------------------|
| 1a. | I permit my sample to be stored and used in future research of neurologic disease at (home institution). | <input type="checkbox"/> 0 No | <input type="checkbox"/> 1 Yes |
| 1b. | I permit my sample to be stored and used in future research at (home institution) to learn about, prevent, or treat other health problems. | <input type="checkbox"/> 0 No | <input type="checkbox"/> 1 Yes |
| 1c. | There is a small chance that some commercial value may result from my sample at the National Cell Repository for Alzheimer's Disease (NCRAD). If that were to happen, I would not be offered a share in any profits. I permit (home institution) to give my sample to researchers at other institutions. | <input type="checkbox"/> 0 No | <input type="checkbox"/> 1 Yes |

Please complete a copy of this form for each relative from whom a specimen was obtained.

Form B3F: Supplemental UPDRS

INSTRUCTIONS: This form is to be completed by the clinician or other trained health professional. For additional clarification and examples, see FTLD Module Coding Guidebook for Follow-up Visit Packet, Form B3F. Check only one box per question.

This form should be completed by a clinician or other trained health professional, based on neurological examination of the subject.

QUESTIONS A1 – A3: For each question in Section A, choose the description that most accurately reflects the subject’s current condition. Select only one box per question.

| | Not to a degree that would justify such a diagnosis | Yes — with asymmetry | | Yes — without major asymmetry | Untestable |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------|----------------------------|----------------------------|-------------------------------|------------|
| | | L>R | R>L | | |
| SECTION A | | | | | |
| A1. Does the subject have limb or torso fasciculations consistent with a diagnosis of spinal muscular atrophy (SMA) or amyotrophic lateral sclerosis (ALS)*? | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | |
| A2. Does the subject have limb weakness and/or hyperreflexia consistent with a diagnosis of primary lateral sclerosis (PLS) or ALS*? | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | |
| A3. Does the subject have bulbar weakness and/or fasciculations consistent with a diagnosis of ALS*? | <input type="checkbox"/> 0 | | | <input type="checkbox"/> 3 | |

Use the following criteria for Questions A1 – A3, 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: | The diagnosis of ALS requires the ABSENCE of: |
| Evidence of lower motor neuron (LMN) degeneration by clinical, electrophysiological or neuropathologic examination; | Electrophysiological or pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration; and |
| Evidence of upper motor neuron (UMN) degeneration by clinical examination; and | Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs. |
| 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. | |

¹Brooks BR, Miller RG, Swash M, Musant TL, for the World Federation of Neurology Research Group on Motor Neuron Diseases. El Escorial revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2000; 1(5):293-299.

Section B, gait disturbances. For each question in Section B, choose the description that most accurately reflects the subject's current condition. Check only one box per question.

| SECTION B Gait disturbances | |
|----------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| B1. Gait disturbance — severity | <input type="checkbox"/> 0 Normal <input type="checkbox"/> 1 Slight alteration in speed or fluidity of gait <input type="checkbox"/> 2 Walks with difficulty but requires no assistance <input type="checkbox"/> 3 Severe disturbance <input type="checkbox"/> 4 Cannot walk at all <input type="checkbox"/> 8 Untestable (SPECIFY REASON): _____ |
| B2. Gait disturbance — type | <input type="checkbox"/> 0 Normal <input type="checkbox"/> 1 Hemiparetic (spastic) <input type="checkbox"/> 2 Foot drop gait (lower motor neuron) <input type="checkbox"/> 3 Ataxic gait <input type="checkbox"/> 4 Parkinsonian gait <input type="checkbox"/> 5 Apractic gait (“magnetic gait”) <input type="checkbox"/> 6 Antalgic gait <input type="checkbox"/> 7 Other gait disorder not listed above (SPECIFY): _____ <input type="checkbox"/> 8 Untestable (SPECIFY REASON): _____ |

The dominant gait disorder should be noted. If there are truly multiple gait disorders, select 7=**Other gait disorder**.

1. **A hemiparetic gait** is typically seen in someone with a stroke or other brain lesion that affects one corticospinal pathway. The paretic limb does not flex at the knee, the foot is plantar flexed, and the leg is circumducted while walking. Often the ipsilateral upper extremity is held in a flexed posture.
2. **A foot-drop gait** is typically seen with weakness of the foot dorsiflexors. It can be bilateral, as in a motor polyneuropathy, or unilateral, as in the case of an isolated peroneal palsy. It could also be seen in motor neuron disease.
3. **An ataxic gait** is seen in someone with midline cerebellar disease, disease of the spinocerebellar pathways, disorders that affect the dorsal columns, or disorders that cause a sensory neuropathy. The gait is typically wide-based. Weaving from side to side may occur.
4. **Parkinsonian gait** is well characterized by a stooped posture, shortened stride, reduced arm swing, and “en bloc” turning, requiring several steps to turn. Note that if Parkinsonian gait is endorsed, Parkinsonian gait disorder (Question 2e on UDS Form B8) should also be selected as present.
5. **Apractic gait.** “Magnetic gait” is a disorder that is associated with normal-pressure hydrocephalus. It consists of a difficulty in initiating gait in the absence of weakness of the legs, while ability is preserved to execute lower-limb motor activity in the sitting position. Parkinsonian gait can resemble an apractic gait, but if there are other signs of Parkinsonism, Parkinsonian gait and not apractic gait should be endorsed.
6. **Antalgic gait.** Antalgic gait refers to alterations in walking due to orthopedic or other musculoskeletal problems that are not primarily neurologic in origin.
7. **Other gait disorder.** Select 7=**another gait disorder** for a gait disorder not listed above, or if there are truly multiple gait disorders, and no dominant disorder can be identified.

Form B9F: Clinical PPA and bvFTD Features

The purpose of this form is to provide determination of clinical features of primary progressive aphasia (PPA) and behavioral variant frontotemporal dementia (bvFTD). This form should be completed by a clinician with experience in evaluating patients with frontotemporal lobar degeneration. **Conclusions should be based on information obtained through subject, co-participant, medical records, observation, imaging, and/or neuropsychological testing.**

Gateway question for primary progressive aphasia (PPA)

- | | |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|
| 1. Is an acquired disorder of language a prominent element of the clinical presentation of the subject? (I.e., at least one of the characteristics described in Questions 2–11 is “Definitely present”.) | <input type="checkbox"/> 0 No (SKIP TO QUESTION 14) <input type="checkbox"/> 1 Yes |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|

For control subjects, select **0=No** and skip to Question 14. Note that Question 12 (root diagnosis of PPA) will be set to **0=No** for all subjects with **0=No** on Question 1.

**TASKS THAT MAY BE USED TO ASSESS SPEECH AND LANGUAGE FUNCTION
IN PRIMARY PROGRESSIVE APHASIA (PPA)**

| Speech/language function | Task | Behavioral measures | Variant in which impaired |
|--------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------|
| Speech production: Grammar | Picture description task; story retelling (e.g., picture aided); constrained-syntax sentence production task | Grammatical structure; mean length of utterance; speech rate; accuracy of content; melody; prosody; specific error types in word selection; articulation | Nonfluent/agrammatic variant |
| Speech production: Motor speech | Motor speech evaluation, including multiple repetitions of multisyllabic words; diadochokinesis of speech articulators; spontaneous speech | Effortfulness; hesitations; presence of apraxia of speech or dysarthria; specific types of speech sound errors; factors that affect articulation (e.g., word length in syllables) | Nonfluent/agrammatic variant |
| Confrontation naming | Single-word retrieval in response to pictures, sounds, foods, and odors | Error rate; delay in naming; factors that affect naming accuracy (e.g., familiar vs unfamiliar items, nouns vs verbs, semantic category); error types (e.g., semantic errors, phonemic errors) | Severe deficit in semantic variant with semantic errors; moderate impairment in logopenic variant with phonemic errors |
| Repetition | Oral repetition of words, pseudowords, phrases, and sentences | Factors that affect repetition accuracy (e.g., predictability of the phrase, sentence length, grammatical complexity); error types | Logopenic variant with phonological errors |
| Sentence comprehension | Matching orally presented sentences to pictures; answering yes/no questions; following directions | Factors that affect comprehension (e.g., grammatical complexity; reversibility of the sentence, e.g., The boy was kicked by the girl vs. The ball was kicked by the girl) | Nonfluent/agrammatic variant, effect of grammatical complexity; logopenic variant, length and frequency effect |
| Single-word comprehension | Word-to-picture matching; Word-to-definition matching; Synonym matching | Factors that affect comprehension (e.g., familiarity; frequency; grammatical word class) | Semantic variant |
| Object/people knowledge | Picture-picture matching; odd-one-out; semantic associations; gesture-object matching; sound-picture matching | Factors that affect object knowledge (e.g., familiarity, semantic category) | Semantic variant |
| Reading/spelling | Lists including regular and irregular word lists, from various word classes, matched for other factors; pseudowords matched to words in length | Factors that affect reading/spelling accuracy (e.g., regularity, frequency, word class); error types (e.g., regularization, phonologically plausible errors; articulatory distortions) | Semantic variant with “regularization” errors; logopenic variant phonologic errors |

Characterizing speech and language symptoms / assigning PPA subtype

Are these features present on the current examination?

Note: Many of these items are also evaluated in the neuropsychological assessment. The responses recorded here should represent the consensus of the clinical and neuropsychological evaluation.

Absent

Questionably present

Definitely present

Not evaluated

2. Poor object naming

(Core diagnostic feature of semantic variant; abnormal in all variants)

0

1

2

9

The subject has difficulty producing names of pictures, objects, sounds, or odors. It can be tested at the bedside with common objects, or with parts of the body.

3. Impoverished word selection / retrieval in spontaneous speech or writing

(Core diagnostic feature of logopenic variant; abnormal in all variants)

0

1

2

9

Spontaneous speech is characterized by hesitations and slow rate, due to difficulty producing content words like nouns and verbs. Typically, there are pauses in speech because of word-finding difficulty.

4. Impaired word comprehension

(Core diagnostic feature of semantic variant; absent in other variants)

0

1

2

9

The subject has difficulties with understanding the meaning of spoken or written single words.

5. Poor object/person knowledge

(Secondary diagnostic feature of semantic variant; absent in other variants)

0

1

2

9

This is different from not being able to name a person (anomia, Question 2) or being unable to define a word (poor comprehension, Question 4). When shown an object such as scissors, the subject may not be able to describe its function. When shown a famous face, the subject may not be able to recognize it as belonging to a politician versus a movie star. When shown three objects (scissors, thimble, paperclip), the subject may not be able to determine which two go together. To determine that this abnormality exists, it is important to ascertain that perceptual abilities are intact.

6. Grammatical simplification or grammatical errors in speech or writing

(Core diagnostic feature of nonfluent/agrammatic variant)

0

1

2

9

In spontaneous speech, either in conversation or in the context of describing a picture, the subject uses short, simple phrases in which grammatical morphemes are omitted (function words, articles, pronouns, inflections).

7. Effortful, halting speech

(Core diagnostic feature of nonfluent/agrammatic variant)

0

1

2

9

Effortful speech is characterized by evident struggles on the subject's part to produce sounds. The speech is often slow, and articulation is likely to be impaired.

| | Absent | Questionably present | Definitely present | Not evaluated |
|---------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 8. Circumlocutory, empty speech (Secondary diagnostic feature of logopenic variant; also present in semantic variant) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |

In spontaneous speech, the subject may use phrases to represent a word, such as, “the thing you use to write with” for “pencil.” The speech may be devoid of substantive words, such as nouns or verbs, which causes the communicative content of the subject’s speech to be low.

| | | | | |
|---------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 9. Speech sound/word errors (paraphasias) (Secondary diagnostic feature of logopenic variant; abnormal in nonfluent/agrammatic variant) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |
|---------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|

Unlike dysarthria, patients may make inconsistent speech sound errors, consisting of distortions, deletions, substitutions, insertions, and/or transpositions of speech sounds. Patients are often aware of these errors.

| | | | | |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 10. Impaired speech repetition (inability to repeat verbatim sentence-length material) (Core diagnostic feature of logopenic variant; present in nonfluent/agrammatic type; absent in semantic variant) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|

Subjects have difficulties repeating sentences that are longer than three or four words. They usually have great difficulty repeating more than two unrelated words (e.g., words that might occur in the context of the learning phase of a brief mental status examination’s memory task).

| | | | | |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 11. Surface dyslexia and dysgraphia — <i>also refer to Word Reading Test from FTLN Neuropsychological Battery</i> (Secondary feature of semantic variant) | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|

Surface dyslexia refers to an impairment in reading and writing words with irregular or atypical relationships between spelling and pronunciation. Patients typically “regularize” such words, so that “sew” is read as /sū/. Surface dyslexia is demonstrated by asking subjects to read irregularly spelled words.

| | | |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------|----------------------------------------------------------------------------------------------------|
| 12. ROOT DIAGNOSIS OF PPA Does the subject have an acquired and progressive difficulty with language consistent with PPA of a neurodegenerative type AND is the language disorder the most prominent deficit at symptom outset and for the initial phase (1–2 years) of the disorder? | <input type="checkbox"/> 0 No (SKIP TO QUESTION 14) | <input type="checkbox"/> 1 Yes — Meets root diagnosis of PPA (PROCEED TO QUESTION 13) |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------|----------------------------------------------------------------------------------------------------|

To meet the PPA diagnosis, all three 2003 *New England Journal of Medicine* core criteria must be met (see table on page 10). A “Yes” response to Question 12 means that the cause is neurodegenerative, that the impairment is progressive, and that the aphasia is the chief impairment in the initial one to two years.

| | Absent | Questionably present | Definitely present | Not evaluated |
|----------------------------------------------------------------------------------------------------------|--------------------------------|---------------------------------------------|--------------------------|-----------------------------|
| 13. Consensus diagnosis of dominant PPA subtype based on clinician and neuropsychologist judgment | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |
| | 1 | 2 | 3 | 4 |
| | PPA, semantic variant (semPPA) | PPA, nonfluent/agrammatic variant (nf/gPPA) | PPA, logopenic variant | PPA not otherwise specified |

For the diagnostic criteria for clinical PPA and its subtypes, see the tables on the following page reproduced from: Gorno-Tempini ML, Hillis AE, Weintraub S, et al. Classification of primary progressive aphasia and its variants. *Neurology* 2011; 76(11):1006-1014.

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.

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

Diagnostic features for NONFLUENT/AGRAMMATIC VARIANT PPA

I. Clinical diagnosis of nonfluent/agrammatic variant PPA

At least one of the following core features must be present:

1. Agrammatism in language production
2. Effortful, halting speech with inconsistent speech sound errors and distortions (apraxia of speech)

At least two of three of the following other features must be present:

1. Impaired comprehension of syntactically complex sentences
2. Spared single-word comprehension
3. Spared object knowledge

II. Imaging-supported nonfluent/agrammatic variant diagnosis

Both of the following criteria must be present:

1. Clinical diagnosis of nonfluent/agrammatic variant PPA
2. Imaging must show one or more of the following results:
 - a. Predominant left posterior fronto-insular atrophy on MRI — or—
 - b. Predominant left posterior fronto-insular hypoperfusion or hypometabolism on SPECT or PET

III. Nonfluent/agrammatic variant PPA with definite pathology

Clinical diagnosis (criterion 1 below) and either Criterion 2 or Criterion 3 must be present:

1. Clinical diagnosis of nonfluent/agrammatic variant PPA
2. Histopathologic evidence of a specific neurodegenerative pathology (e.g., FTLT-tau, FTLT-TDP, AD, other)
3. Presence of a known pathogenic mutation

Diagnostic criteria for SEMANTIC VARIANT PPA

I. Clinical diagnosis of semantic variant PPA

Both of the following core features must be present:

1. Impaired confrontation naming
2. Impaired single-word comprehension

At least three of the following other diagnostic features must be present:

1. Impaired object knowledge, particularly for low-frequency or low-familiarity items
2. Surface dyslexia or dysgraphia
3. Spared repetition
4. Spared speech production (grammar and motor speech)

II. Imaging-supported semantic variant PPA diagnosis

Both of the following criteria must be present:

1. Clinical diagnosis of semantic variant PPA
2. Imaging must show one or more of the following results:
 - a. Predominant anterior temporal lobe atrophy
 - b. Predominant anterior temporal lobe hypoperfusion or hypometabolism on SPECT or PET

III. Semantic variant PPA with definite pathology

Clinical diagnosis (criterion 1 below) and either Criterion 2 or Criterion 3 must be present:

1. Clinical diagnosis of semantic variant PPA
2. Histopathologic evidence of a specific neurodegenerative pathology (e.g., FTLT-tau, FTLT-TDP, AD, other)
3. Presence of a known pathogenic mutation

Abbreviations: AD = Alzheimer's disease; FTLT = frontotemporal lobar degeneration; PPA = primary progressive aphasia

DIAGNOSTIC CRITERIA FOR LOGOPENIC VARIANT PPA

I. Clinical diagnosis of logopenic variant PPA

Both of the following core features must be present:

1. Impaired single-word retrieval in spontaneous speech and naming
2. Impaired repetition of sentences and phrases

At least three of the following other features must be present:

1. Speech (phonologic) errors in spontaneous speech and naming
2. Spared single-word comprehension and object knowledge
3. Spared motor speech
4. Absence of frank agrammatism

II. Imaging-supported logopenic variant diagnosis

Both criteria must be present:

1. Clinical diagnosis of logopenic variant PPA
2. Imaging must show at least one of the following results:
 - a. Predominant left posterior perisylvian or parietal atrophy on MRI
 - b. Predominant left posterior perisylvian or parietal hypoperfusion or hypometabolism on SPECT or PET

III. Logopenic variant PPA with definite pathology

Clinical diagnosis (criterion 1 below) and either Criterion 2 or Criterion 3 must be present:

1. Clinical diagnosis of logopenic variant PPA
2. Histopathologic evidence of a specific neurodegenerative pathology (e.g. AD, FTLT-tau, FTLT-TDP, other)
3. Presence of a known pathogenic mutation

Abbreviations: AD = Alzheimer's disease; FTLT = frontotemporal lobar degeneration; PPA = primary progressive aphasia

Gateway question for behavioral variant frontotemporal dementia (bvFTD)

14. **Are acquired alterations in behavior, personality, or comporment important elements in the clinical presentation of the subject? (I.e., at least one of the characteristics described in Questions 15–21 is “Definitely present”.)**
- 0 No (SKIP TO QUESTION 23)
- 1 Yes

bvFTD gateway question: Question 14 asks the clinician whether there are prominent changes in behavior, personality, or comporment that could justify a more detailed description of those abnormalities, which is obtained with Questions 15–20. Question 14 does not constitute a diagnosis but is only a means for determining whether the clinician completes the detailed assessment of behaviors or skips it.

For control subjects, select **0=No**, and skip to Question 23.

Note that Question 22, clinical bvFTD syndrome, will be set to **0=Does not meet criteria for bvFTD** for all subjects with **0=No** on Question 14.

QUESTIONS 15–21: These six domains are drawn directly from: Rascovsky, K, Hodges JR, Knopman D, et al. Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia. *Brain* 2011; 139(9):2456-2477. The diagnosis of Possible bvFTD is based on personality, social comporment, and cognitive features that distinguish bvFTD from other conditions. While it is important to interpret diagnostic features of a case in the clinical and cultural context, ratings of behavioral features can be difficult and potentially open to subjective bias. As such, ratings should be based on overt behaviors, as opposed to inferences about a subject’s cognitive or emotional state. As these are questions based on the history, the queries and responses should cover the prior three years.

| Characterizing symptoms of bvFTD | | | | |
|-------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| Have the following symptoms/behaviors been prominent, persistent, and recurrent in (approximately) the past three years? | Absent | Questionably present | Definitely present | Not evaluated |
| 15. Disinhibition Socially inappropriate behavior; loss of manners or decorum; impulsive, rash, or careless actions | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |

Early behavioral disinhibition is a hallmark feature of the bvFTD clinical syndrome. This does not include behaviors driven by hallucinations and paranoia. Disinhibition may present as one of the following:

- a. Socially inappropriate behavior.** Examples of behaviors that violate social norms include inappropriately approaching, touching, or kissing strangers; verbal or physical aggression; public nudity or urination; inappropriate sexual acts; and criminal behavior (such as theft or shoplifting).
- b. Loss of manners or decorum.** This category includes a range of behaviors that violate social graces, such as inappropriate laughter, cursing or loudness, offensive jokes or opinions, or crude or sexually explicit remarks. Patients may also display a general lack of etiquette (e.g., failing to wait in line, eating with mouth open), loss of respect for interpersonal space, and a lack of response to social cues (e.g., patient will continue talking despite others’ attempts to end a conversation). Some bvFTD patients exhibit poor hygiene/grooming (e.g., wearing malodorous, stained, torn or inappropriate clothing) or impolite physical behaviors (e.g., flatulence, scratching or fondling private parts, picking teeth, belching, or spitting).
- c. Impulsive, rash, or careless actions.** Impulsive behaviors may include reckless driving, new-onset gambling, stealing (usually food or shiny objects), buying/selling objects without regard for consequences, or indiscriminate sharing of personal information (e.g., credit card information, Social Security number).

| | | | | |
|-----------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 16. Apathy or inertia Loss of interest, drive, and motivation; decreased initiation of behavior | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |
|-----------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|

Early apathy or inertia. Apathy/inertia is the most common initial symptom in bvFTD. It is important to distinguish this from the poor initiation of activities that can be seen in depression. One of the following symptoms should be present:

- a. Apathy.** Apathy is defined as a loss of motivation, drive or interest. It can manifest as passivity or lack of spontaneity. The patient may lack initiative and cease to engage in important or previously rewarding activities (e.g., job, hobbies).
- b. Inertia.** Inertia refers to decreased initiation of behavior (i.e., the patient requires prompts or cues to initiate or continue routine activities). For example, it may be reported that a patient requires specific directives to start and finish brushing his teeth, or that a patient no longer starts or sustains conversation.

Characterizing symptoms of bvFTD

Have the following symptoms/behaviors been prominent, persistent, and recurrent in (approximately) the past three years?

| | Absent | Questionably present | Definitely present | Not evaluated |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 17. Loss of sympathy / empathy Diminished response to other people's needs or feelings; diminished social interest, interrelatedness, or personal warmth | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |

Early loss of sympathy or empathy. Loss of empathy refers to an inability to read the emotional expressions of others or imagine their experiences. It is a common feature at initial presentation, and is often coupled with indifference and a general decrease in social engagement. In everyday life, loss of sympathy or empathy may present as one of the following:

- a. Diminished responsiveness to other people's needs and feelings.** A positive rating on this feature should be based on specific examples that reflect a lack of understanding or indifference to the feelings of others (e.g., hurtful comments or inexplicable disregard for others' pain or distress).
- b. Diminished social interest, interrelatedness or personal warmth.** This feature refers to a more general decline in social engagement, with emotional detachment, coldness, lack of eye contact, etc. Relatives and friends might experience the patient as uncharacteristically distant (e.g., no longer touches, hugs, or seeks out their company).

| | | | | |
|----------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 18. Ritualistic / compulsive behavior Simple repetitive movements or complex compulsive or ritualistic behaviors | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |
|----------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|

Early perseverative, stereotyped, or compulsive/ritualistic behavior. A positive rating on this feature can occur if the patient exhibits any one of the following:

- a. Simple repetitive movements.** These movements include tapping, clapping, rubbing, scratching, picking at skin or clothing, humming, rocking, throat clearing, pursing of lips or lip smacking.
- b. Complex, compulsive/ritualistic behaviors.** Examples include counting and cleaning rituals, collecting or hoarding, checking, repetitive trips to the bathroom (without need), ordering objects, and walking fixed routes. This can encompass a complex belief system such as a radical change in religious or political beliefs. Pacing (without a compulsive quality) should not be included, as it can occur in other primary dementias or as a psychotropic medication effect.
- c. Stereotypy of speech.** These are single words, phrases or entire themes or stories that the patient habitually repeats, despite their lack of communicative value.

Characterizing symptoms of bvFTD

Have the following symptoms/behaviors been prominent, persistent, and recurrent in (approximately) the past three years?

| | Absent | Questionably present | Definitely present | Not evaluated |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 19. Hyperorality and appetite changes Altered food preferences, binge eating, increased consumption of alcohol or cigarettes, oral exploration or consumption of inedible objects | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 9 |

Changes in dietary and eating behavior are common manifestations of bvFTD, and can range from altered food preferences to oral exploration of inedible objects. This feature can present as one of the following symptoms:

- a. **Altered food preferences.** In the context of bvFTD, this change in food habits usually presents as carbohydrate cravings (particularly sweets) or food fads (i.e., rigid, stereotyped, or idiosyncratic food preferences).
- b. **Binge eating, increased consumption of alcohol or cigarettes.** Patients consume excessive amounts of food and continue to eat despite (in some cases) acknowledging satiety. Some patients exhibit new, resumed, or compulsive smoking or ingestion of alcohol.
- c. **Oral exploration or consumption of inedible objects.** In extreme cases, hyperorality may manifest as oral exploration, chewing, or ingestion of inedible objects. Hyperorality is a feature consistent with Klüver-Bucy syndrome.

20. **Changes on neuropsychological testing consistent with bvFTD** (refer to neuropsychological evaluation and neuropsychologist's impression)

0 1 2 9

This item requires review of the neuropsychological testing. The neuropsychological profile supportive of a diagnosis of bvFTD should exhibit executive/generation deficits with relative sparing of memory and visuospatial functions. All of the following features must be present: 1.) deficits in executive tasks; 2.) relative sparing of episodic memory; 3.) relative sparing of visuospatial skills. Note that this information could be obtained in the present or could be obtained from historical records.

21. **Impaired daily functioning**
Are these alterations in behavior, personality, or comporment the principal cause of impaired daily living activities?

0 1 2 9

This question asks whether the patient exhibits significant functional decline as per caregiver report. This criterion must be fulfilled — a rating of definite impairment — for a diagnosis of dementia generally and is a requirement for the diagnosis of probable bvFTD.

22. **Does the subject meet the criteria for clinical probable* or possible** bvFTD syndrome — in other words, are features described in Questions 15–20 the most prominent parts of the clinical picture now or initially?”**

*PROBABLE: Meets three of the above criteria in Questions 15–20 and: (1) has impaired daily functioning (Question 21 = Definitely Present); and (2) has imaging consistent with bvFTD.

**POSSIBLE: Meets three of the above criteria in Questions 15–20 and either: (1) is not functionally impaired (i.e., Question 21= Absent) or (2) does not have imaging consistent with bvFTD.

0 0 = Does not meet criteria for bvFTD: either subject meets <3 of the features described in Questions 15–20, OR another diagnosis is suggested by prominent features not covered by Questions 15–20

1 Probable bvFTD

2 Meets criteria for possible bvFTD and has impaired daily functioning but without evidence of diagnostic imaging

3 Meets criteria for possible bvFTD (with or without evidence of diagnostic imaging), but daily functioning is not significantly impaired

Question 22, the formal diagnostic question for bvFTD. The response is based on the information in Questions 15–21. The diagnosis also depends upon imaging findings. There are three explicit exclusions:

- 1.) Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders;
- 2.) Behavioral disturbance is better accounted for by a psychiatric diagnosis;
- 3.) Biomarkers strongly indicative of Alzheimer’s disease or other neurodegenerative process.

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 = behavioural variant FTD*

QUESTIONS 23 – 25. Self-explanatory questions regarding whether an electromyogram (EMG) was performed. Check only one box per question.

| | No | Yes | Uncertain |
|----------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 23. Was an electromyogram (EMG) performed at this visit? If answer is "1 (Yes)", SKIP TO QUESTION 25. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |
| 24. Has an EMG been performed in the past year? If answer is "0 (No)", SKIP TO QUESTION 26. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |
| 25. If an EMG was performed, did it show evidence of motor neuron disease? | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

Select **9=Uncertain** only if evidence of motor neuron disease is not conclusive.

If subject has only one diagnosis (either PPA or bvFTD), then END FORM HERE.

26. For subjects with a diagnosis of both PPA and bvFTD, which diagnosis appeared first?
- 1 bvFTD
 - 2 PPA, semantic variant
 - 3 PPA, nonfluent/agrammatic variant
 - 4 PPA, logopenic variant
 - 5 PPA not otherwise specified
 - 9 Unknown

Question 26 clarifies the order of appearance in cases where more than one of the syndromes is present. For subjects with a diagnosis of PPA and bvFTD, indicate which diagnosis appeared first. Do not answer this question if only one diagnosis is present.

Select **9=Unknown** only if the order of appearance is unknown.

Form C1F: Neuropsychological Battery Summary Scores

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

It is intended that the tests be administered in the order in which they appear, even if they were previously administered at a recent clinic screening. This is necessary in order to standardize test administration among Centers. It is therefore suggested that the FTLD Neuropsychological Battery be administered in its entirety after the Uniform Data Set (UDS) neuropsychological battery and either before or after the administration of other tests commonly used by the Center.

The instructions provided within the *FTLD Module— Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* should be closely followed at all times since these instructions may be different from Center-specific protocols that may already be in place.

Videotaping is recommended if the examiner is not familiar with language disorders. The tapes can then be viewed by clinicians who are experienced in language disorders, and these clinicians can assist with scoring. Alternatively, examiners should write the subject's response verbatim and seek the help of a clinician at their Center who is familiar with language disorders. If the answer is still unclear after these steps are taken, please contact NACC, which will forward your questions to the FTLD Neuropsychology work group.

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. Thus, if a participant notices an error after drawing the complete Benson complex figure, for example, do not allow the correction. However, if when the participant starts to draw an erroneous line and immediately self-corrects, permit the correction.

INSTRUCTIONS: This form is to be completed by ADC or clinic staff. For test administration and scoring, see FTLD Module Coding Guidebook for Follow-up Visit Packet, Form C1F. Time to completion of C1F and C2F tests should be reported at the end of Form C3F.

KEY: If the subject cannot complete any of the following exams, please give the reason by entering one of the following codes:

95 = Physical problem 96 = Cognitive/behavior problem 97 = Other problem 98 = Verbal refusal

1. Word Reading Test — Regular and Irregular Words

REGULAR

1a. Total completely accurate words (0-15, ____ ____
(If test not completed, enter reason code, 95-98, and SKIP TO QUESTION 2a) 95-98)

1b. Total semantically related inaccurate words (0-15) ____ ____

1c. Total other phonologically related words or nonword errors (0-15) ____ ____

IRREGULAR

1d. Total completely accurate words (0-15) ____ ____

1e. Total semantically related inaccurate words (0-15) ____ ____

1f. Total words that are “regularized” (read using “phonics,” e.g., *sew* read as *sue*) (0-15) ____ ____

1g. Total other phonologically related words or nonword errors (0-15) ____ ____

This is a test of word reading that includes regularly spelled and irregularly spelled words. In this test, a subject is asked to read out loud from the regular and irregularly spelled word lists. The primary measures of performance are the total completely accurate words (both regular and irregular).

Review *FTLD Module — Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet provided and enter the appropriate scores here.

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2. Semantic Word-picture Matching Test

2a. Total correct word-picture matches (0-20, ____ ____
(If test not completed, enter reason code, 95-98, and SKIP TO QUESTION 3a) 95-98)

This test evaluates spoken word recognition and assesses the frequency of semantic errors in word comprehension. The stimuli consist of five four-picture displays, each of which includes pictures of four objects that are semantically related. These five displays are each presented four times (once for each picture as the target), for a total of 20 trials.

Review *FTLD Module — Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet provided, and enter the appropriate score here.

Rogalsky C, Love T, Driscoll D, Anderson SW, and Hickok G. *Are mirror neurons the basis of speech perception? Evidence from five cases with damage to the purported human mirror system.* *Neurocase.* 2011;17(2):178-87. Reproduced by permission.

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KEY: If the subject cannot complete any of the following exams, please give the reason by entering one of the following codes:

95 = Physical problem **96 = Cognitive/behavior problem** **97 = Other problem** **98 = Verbal refusal**

3. Semantic Associates Test

| | |
|-----------------------------------------------------------------------------------------------------------------------------|-------------------------|
| 3a. Total correct animal associations (If test not completed, enter reason code, 95–98, and SKIP TO QUESTION 4a) | (0–8, 95–98) ___ ___ |
| 3b. Total correct tool associations | (0–8) ___ ___ |
| 3c. Sum of all correct associations (Semantic Associates Test total score) | (0–16) ___ ___ |

This is a test of knowledge of the meaning of objects. In this test, a subject reviews pairs of pictures and is instructed to select those that depict related objects. The primary measure of performance is the sum of all correct associations (Semantic Associates Test total score).

Review *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet provided and enter the appropriate scores here.

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4. Northwestern Anagram Test — Short Form

| | |
|-------------------------------------------------------------------------------------------------------------------------|-------------------------|
| 4a. Correct subject who-questions (If test not completed, enter reason code, 95–98, and SKIP TO QUESTION 5a) | (0–5, 95–98) ___ ___ |
| 4b. Correct object who-questions | (0–5) ___ ___ |
| 4c. Total score: sum of all correct questions | (0–10) ___ ___ |

This is a test of grammatical knowledge. In this test, the subject is shown pictures and is then asked to assemble a sentence describing the pictures using printed words that are provided. The primary measure of performance is the total score (sum of all correct questions).

Review *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet and enter the appropriate scores here.

From the Northwestern Anagram Test (Short Form, 2012), Cynthia K. Thompson, Sandra Weintraub, and Marsel Mesulam (<https://flintbox.com/public/project/19927>); further copying or distribution is forbidden without authors' permission. Forms created as part of the FTLD Module to the Uniform Data Set of the National Alzheimer's Coordinating Center.

KEY: If the subject cannot complete any of the following exams, please give the reason by entering one of the following codes:

95 = Physical problem 96 = Cognitive/behavior problem 97 = Other problem 98 = Verbal refusal

5. Sentence Repetition Test

| | |
|-----------------------------------------------------------------------------------------------------------------------------------|---------------------------|
| 5a. Number of completely accurate sentences (If test not completed, enter reason code, 95–98, and SKIP TO QUESTION 6a) | (0–5, 95–98) ___ ___ |
| 5b. Total number of words omitted from sentences | (0–37) ___ ___ |
| 5c. Total number of semantically related or unrelated incorrect real words | (0–20) ___ ___ |
| 5d. Total number of phonologically related words or nonword errors | (0–20) ___ ___ |

This is a test of oral repetition of sentence-length utterances. In this test, the clinician reads a sentence out loud to the subject. The subject then repeats the sentence verbatim. Correct sentences, omitted words and semantic errors are recorded for scoring purposes. The primary measure of performance is the number of completely accurate sentences.

Review *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet provided and enter the appropriate scores here.

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6. Noun and Verb Naming Subtests

| | |
|---------------------------------------------------------------------------------------------------------------|-------------------------------------------------------|
| 6a. Total nouns correct (If test not completed, enter reason code, 95–98, and SKIP TO QUESTION 7a) | (0–16, 95–98) ___ ___ |
| 6b. Total verbs correct | (0–16) ___ ___ |
| 6c. Noun-to-verb ratio (total nouns correct / total verbs correct) | (88.88 = Cannot be calculated) ___ ___ . ___ ___ |

This is a test of confrontation naming of objects and actions. In this test, the subject is shown pictures of objects or things, as well as pictures of people doing various actions. The subject is then asked to name each picture as quickly and as accurately as possible. The primary measure of performance is the noun-to-verb ratio (total nouns correct/ total verbs correct).

If either the noun or verb score is zero, the noun-to-verb ratio should not be calculated. In this case, please enter “88.88”.

Review *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet and enter the appropriate scores here.

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KEY: If the subject cannot complete any of the following exams, please give the reason by entering one of the following codes:

95 = Physical problem

96 = Cognitive/behavior problem

97 = Other problem

98 = Verbal refusal

7. Sentence Reading Test

| | | |
|-----------------------------------------------------------------------------------------------------------------------------|--------------|---------|
| 7a. Number of completely accurate sentences (If test not completed, enter reason code, 95–98, and END FORM HERE) | (0–5, 95–98) | ___ ___ |
| 7b. Total number of words omitted from sentence | (0–37) | ___ ___ |
| 7c. Total number of semantically related or unrelated incorrect real words | (0–20) | ___ ___ |
| 7d. Total number of phonologically related words or nonword errors | (0–20) | ___ ___ |

This is a test of sentence reading. In this test, the subject is given a sheet of paper with five short sentences and is asked to read the sentences out loud. The primary measure of performance is the number of completely accurate sentences.

Review *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*, complete the worksheet and enter the appropriate scores here.

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Form C2F: Social Norms Questionnaire¹

The intent of the Social Norms Questionnaire is to determine how well subjects can understand and identify social boundaries that are part of mainstream culture in the United States. Ask the subject to check the most accurate response for each item below. Tell the subject that “don’t know” and “not applicable” are not allowable responses for any item. See *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* for more details on this form.

INSTRUCTIONS FOR THE CENTER

This questionnaire is designed to be completed **by the subject in the presence of a qualified psychologist or psychometrist** as part of a face-to-face battery of tests. The examiner should read and explain the instructions to the subject, then ask the subject to complete the questionnaire. If the subject asks for clarification of the procedure or questions, it is acceptable for the examiner to discuss the questionnaire with him or her. Tell the subject that “Don’t know” and “Not applicable” are not allowable responses for any item. While it is permissible to help a cognitively impaired subject understand and complete the questionnaire (e.g., reading the questions out loud, or marking their response for them), the examiner should ensure that they merely help the subject understand a question (e.g., “Do you think it’s OK to cut in line if you are in a hurry?”), but not help them to formulate their response. In this way, if the subject asks what they should answer, it would be permissible to respond with prompts such as, “It’s up to you. Answer whatever you think is best. It’s OK to guess if you’re not sure.”).

KEY: If the subject is so impaired as to make administration of this questionnaire impossible, please give the reason by checking one of the following reason codes in the “FOR CLINIC USE ONLY” section and skip the remaining data elements.

95=Physical problem 96=Cognitive/behavior problem 97=Other problem 98=Verbal refusal

If the subject completes some but not all of the questionnaire, items that are missing should be left blank, and all affected summary and total scores should be entered as 88 or 88.88, as appropriate.

Instructions: Following is a list of behaviors that a person might engage in. Please decide whether or not it would be socially acceptable and appropriate to do these things in the mainstream culture of the United States and answer yes or no to each. Think about these questions as if they were occurring in front of or with a stranger or acquaintance, NOT a close friend or family member.

WOULD IT BE SOCIALLY ACCEPTABLE TO:

| | | |
|----------------------------------------------------|-------------------------------|--------------------------------|
| 1. Tell a stranger you don’t like their hairstyle? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 2. Spit on the floor? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 3. Blow your nose in public? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 4. Ask a coworker their age? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 5. Cry during a movie at the theater? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |

| | | |
|-------------------------------------------------------------|-------------------------------|--------------------------------|
| 6. Cut in line if you are in a hurry? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 7. Laugh when you yourself trip and fall? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 8. Eat pasta with your fingers? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 9. Tell a coworker your age? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 10. Tell someone your opinion of a movie they haven't seen? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 11. Laugh when someone else trips and falls? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 12. Wear the same shirt every day? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 13. Keep money you find on the sidewalk? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 14. Pick your nose in public? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 15. Tell a coworker you think they are overweight? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 16. Eat ribs with your fingers? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 17. Tell a stranger you like their hairstyle? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 18. Wear the same shirt twice in two weeks? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 19. Tell someone the ending of a movie they haven't seen? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 20. Hug a stranger without asking first? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 21. Talk out loud during a movie at the theater? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |
| 22. Tell a co-worker you think they have lost weight? | <input type="checkbox"/> 0 NO | <input type="checkbox"/> 1 YES |

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FOR CLINIC USE ONLY. Note: Calculation of the four summary scores below is OPTIONAL.

23. SNQ22 Total Score (0–22): _____

24. Break Score (0–12): _____

25. Overadhere Score (0–10): _____

26. Yes-No Ratio Score (0–22): _____ . _____

SCORING INSTRUCTIONS FOR FORM C2F

The instructions below are for deriving the summary scores (Items 23–25) and Yes/No Ratio Score (Item 26).

However, calculation of these scores is OPTIONAL, as these are automatically calculated and entered upon submission of the form.

Note that the coding scheme on the form (**No=0** and **Yes=1**) is used only for recording item-level data and does not play a role in deriving the summary scores. Instead, item responses are scored as correct or incorrect based on the scoring key (see next page), and a coding scheme of **Correct=0** and **Incorrect=1** is applied. Following are the formulas for the summary scores:

- **SNQ22 Total Score (optional)** is calculated as [22 minus (the sum of items 1 to 22)], ranging from 0 to 22 (higher scores reflecting better performance). If an item is missing, the total score should not be calculated. In this case, enter 88.
- **Break Score (optional)** is the total number of errors made in the direction of breaking a social norm, and is calculated as (sum of Items 1, 2, 4, 6, 8, 11, 12, 14, 15, 19, 20, 21), ranging from 0 to 12 (higher scores reflecting more errors). If an item is missing, the total score should not be calculated. In this case, enter 88.
- **Overadhere Score (optional)** is the total number of errors made in the direction of overadherence to a perceived social norm, and is calculated as (sum of Items 3, 5, 7, 9, 10, 13, 16, 17, 18, 22), ranging from 0 to 10 (higher scores reflecting more errors). If an item is missing, the total score should not be calculated. In this case, enter 88.

Yes/No Ratio Score (optional). In cases where it is unclear whether the subject's cognitive or behavioral deficits have caused them to answer in a stimulus-bound or otherwise meaningless manner, the validity of subject performance can also be measured by determining the ratio of Yes to No responses. The Yes/No Ratio Score, ranging from 0 to 22, can be calculated by counting the number of items to which the subject responded "Yes" and dividing by the number of items to which the subject responded "No." If this score is greater than or equal to 5, or is less than 0.3, please consider whether the subject was too impaired to fill out the form or answered the questions in a meaningless way. If the subject's answers are deemed valid, then please submit the data as is. However, if there is reason to suspect that the values are not valid, please select the appropriate reason code in the "For clinic use only" section and leave the rest of the form blank.

If the subject answered the C2F questions all yes or all no, then these answers are considered invalid. Please select the most appropriate reason code in the "For clinic use only" section and leave the rest of the form blank.

SCORING KEY

Social Norms Questionnaire (SNQ22)

| | CORRECT RESPONSE |
|-------------------------------------------------------------|------------------|
| 1. Tell a stranger you don't like their hairstyle? | NO |
| 2. Spit on the floor? | NO |
| 3. Blow your nose in public? | YES |
| 4. Ask a coworker their age? | NO |
| 5. Cry during a movie at the theater? | YES |
| 6. Cut in line if you are in a hurry? | NO |
| 7. Laugh when you yourself trip and fall? | YES |
| 8. Eat pasta with your fingers? | NO |
| 9. Tell a coworker your age? | YES |
| 10. Tell someone your opinion of a movie they haven't seen? | YES |
| 11. Laugh when someone else trips and falls? | NO |
| 12. Wear the same shirt every day? | NO |
| 13. Keep money you find on the sidewalk? | YES |
| 14. Pick your nose in public? | NO |
| 15. Tell a coworker you think they are overweight? | NO |
| 16. Eat ribs with your fingers? | YES |
| 17. Tell a stranger you like their hairstyle? | YES |
| 18. Wear the same shirt twice in two weeks? | YES |
| 19. Tell someone the ending of a movie they haven't seen? | NO |
| 20. Hug a stranger without asking first? | NO |
| 21. Talk out loud during a movie at the theater? | NO |
| 22. Tell a coworker you think they have lost weight? | YES |

Form C3F: Social Behavior Observer Checklist¹

The intent of the Social Behavior Observer Checklist is to assist clinicians with the recognition of distinct patterns of spontaneous social behaviors. Check the most accurate response for each item below. “Don’t know” and “not applicable” are not allowable responses for any item. See *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* for more details on this form.

INSTRUCTIONS: This form is to be completed by the examiner who administered the neuro-psychological battery to the subject. For additional clarification and examples, see *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F*. Check only one box per question.

Directions: Immediately after the end of your evaluation of the subject, please rate his/her behavior during the time he/she was with you. Use the scales for both the main descriptors (i.e., 1, 2, 3 ...) and the behavior counts (a., b., c. ...) and complete all items.

Your descriptor ratings and behavior counts for the same item can be independent. You may describe the subject as having a particular characteristic on a main descriptor, even if you endorse “never” for all of the behavior counts for that item, or vice versa.

| | | | | |
|----------------------------------------------------------------------------------------------|----------------------------------------------|------------------------------------------------|----------------------------------------------|--------------------------------------------|
| 1. Was overly self-conscious / embarrassed for self: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Spontaneously mentioned that he/she was performing badly | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Made other self-deprecatory comments | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Showed emotional distress over his/her performance / cognitive abilities | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 2. Showed too little self-consciousness / embarrassment for self: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Disrobed immodestly (took off shoes, belt, pants, etc.; lifted shirt, etc.) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Engaged in belching or flatulence, or picked nose without apology | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Giggled or otherwise made silly, childish comment or noise | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 3. Insensitive to others’ embarrassment or privacy: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Insulted or made a negative comment about examiner | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Made an embarrassing comment about examiner | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Made an inappropriate or embarrassing joke | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 4. Failed to adapt / defer to structure of testing situation established by examiner: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Resisted redirection while engaging in a verbal monologue | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Interrupted examiner | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |

| | | | | |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------|------------------------------------------------|----------------------------------------------|--------------------------------------------|
| c. Verbalized a desire to leave the evaluation prematurely | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| d. Physically attempted to leave the evaluation prematurely | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| e. Failed to maintain topic of discussion, initiated tangent | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| f. Demanded that test protocol be broken for him/her (e.g., insisted on completing an item after being told to stop, tried to cheat, tried to turn page to advance to next item against examiner's expressed wishes, etc.) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 5. Was preoccupied with time or kept a strict timetable: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Reminded examiner what time evaluation had to be finished | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 6. Acted overly dependent: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Mentioned caregiver's absence or asked when caregiver would return | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Asked for feedback on performance | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Showed frustration when examiner would not provide explicit feedback | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 7. Was anxious: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Mentioned being nervous about testing / performance anxiety | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Mentioned being nervous about diagnosis or prognosis | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 8. Was stimulus-bound: | <input type="checkbox"/> 0 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Made stimulus-bound error on testing | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Picked up object on desk unnecessarily | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Circumstantial speech; overly focused on details, overly lengthy | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 9. Was perseverative: | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Repeated previous answer on testing | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Repeated an anecdote | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 10. Showed decreased initiation: | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Began response in a notably delayed manner (not due to general slowing) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Required additional verbal prompting to initiate task | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |

| | | | | |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------|-----------------------------------------|---------------------------------------|-------------------------------------|
| 11. Showed fluctuating level of cognitive ability through sessions regardless of complexity of material (e.g., was coherent at times and had notable difficulty understanding at other times): | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Lost task set / forgot instructions after performing task correctly | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Repeated rules to self multiple times during task | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Lost train of thought during conversation or response (demonstrated thought blocking) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 12. Was overly disclosing or inappropriately familiar: | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| a. Spontaneously revealed inappropriately personal information concerning self (only) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| b. Spontaneously revealed inappropriately personal information concerning a relative or friend (can also involve self) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| c. Stood or leaned too close to examiner (noticeably entered examiner's personal space) | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| d. Touched examiner | <input type="checkbox"/> 0 Never | <input type="checkbox"/> 1 Once | <input type="checkbox"/> 2 2-3x | <input type="checkbox"/> 3 4+ |
| 13. Showed diminished social / emotional engagement: | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| 14. Showed exaggerated / labile emotional reactivity: | <input type="checkbox"/> 1 Not at all | <input type="checkbox"/> 1 A little bit | <input type="checkbox"/> 2 Moderately | <input type="checkbox"/> 3 Severely |
| 15. DESCRIPTOR TOTAL SCORE (0–42): | _____ | | | |
| 16. CHECKLIST (BEHAVIOR) SCORE (0–105): | _____ | | | |

17. LENGTH OF THE ENTIRE FTLD NEUROPSYCHOLOGICAL TESTING SESSION:

Record in minutes the approximate length of the testing session upon which these checklist responses were based. This should include, at minimum, time spent on all _____ tests in the FTLD neuropsychological battery (all tests recorded on Form C1F, plus Form C2F), as well as time spent administering any other neuropsychological tests.

Record **approximately** how long it took the subject to complete the testing session (i.e., C2F and all tests reported on C1F).

SCORING INSTRUCTIONS FOR FORM C3F

- Each descriptor and checklist item represents a separate score, ranging from 0 to 3, which can be analyzed to provide independently meaningful data.
- Also, scores for all descriptors 1–14 can be summed to derive the **Descriptor Total Score** (range: 0–42)
- All 35 checklist (behavior) items can be summed to derive the **Checklist (Behavior) Total Score** (range: 0–105). Higher scores are interpreted as reflecting a greater degree of behavioral disturbance.

Form C4F: Behavioral Inhibition Scale¹

The intent of the Behavior Inhibition Scale (BIS) is to assess the subject's current behavioral tendencies, in particular inhibitory and excitatory tendencies. Ask the informant to check the most accurate response for each item below. Tell the informant that "don't know" and "not applicable" are not allowable responses for any item. See *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* for more details on this questionnaire.

INSTRUCTIONS FOR THE CENTER

This questionnaire is designed to be completed **independently by the informant**, who will be describing the subject's current typical behavior. This form may be handed to the informant for completion by him- or herself at any time during the study visit. Tell the informant that "Don't know" and "Not applicable" are not allowable responses for any item. If the informant asks for clarification of questions, it is acceptable for a qualified psychologist or psychometrist to discuss the questionnaire with him or her. However, if the informant completes this questionnaire collaboratively with the clinician, either face-to-face or via telephone, **you must inform NACC of this change in protocol** by checking the appropriate box in the gray "FOR CLINIC USE ONLY" area at the top of the questionnaire.

Before the informant leaves, clinic staff should make sure that all questions were completed by the informant (i.e., none was left blank) by discussing the missing item with the informant and encouraging them to provide a response. If this is not done and it is later noticed that some items were missed by the informant, clinic staff should call the informant as soon as possible so that the missing items can be completed by phone. In this case, the questionnaire is not considered to have been completed independently by the informant. In the shaded area at the top of the form, the appropriate response would therefore be, "This questionnaire was completed via telephone interview of informant by clinic staff."

If there are still missing items, these items should be left blank, and "88" should be entered for the total score.

THIS QUESTIONNAIRE WAS COMPLETED:

- 0 Independently by co-participant, as described in "Instructions to the Center"
- 1 Via in-person interview of co-participant by clinic staff
- 2 Via telephone interview of co-participant by clinic staff

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, select **2=Via telephone interview of informant by clinic staff**. Select only one method.

INSTRUCTIONS: Indicate how well each statement describes the subject's **CURRENT** behavior. There are no right or wrong answers; we just want to get your impression of how you think the subject typically behaves.

If you have questions about how to complete this questionnaire, please ask a staff member, and they will be happy to help you.

| | Strongly Disagree (1) | Disagree (2) | Agree (3) | Strongly Agree (4) |
|------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 1. If the subject thinks something unpleasant is going to happen, he/she usually gets pretty "worked up." | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 2. The subject worries about making mistakes. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 3. Criticism or scolding hurts the subject quite a bit. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 4. The subject feels pretty worried or upset when he/she thinks somebody is angry at him/her. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 5. Even if something bad is about to happen to the subject, he/she rarely experiences fear or nervousness. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 6. The subject feels worried when he/she thinks he/she has done poorly at something. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |
| 7. The subject has very few fears compared to his/her friends. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 |

¹ Copyright © 1994 by the American Psychological Association. Adapted with permission. The official citation that should be used in referencing this material is Table 1 (adapted), p. 323, from Carver C. S., & White, T. L. (1994). Behavioral inhibition, behavioral activation, and affective responses to impending reward and punishment: The BIS/BAS Scales. *Journal of Personality and Social Psychology*, 67(2), 319-333. doi:10.1037/0022-3514.67.2.319. No further reproduction or distribution is permitted without written permission from the American Psychological Association.

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8. BIS Total Score (7–28): ___ ___

SCORING INSTRUCTIONS FOR FORM C4F

Each item yields a score from 1 to 4.

The **BIS Total Score** is calculated as follows; please note that the formula below performs the required reverse scoring of items 5 and 7:

$$\text{BIS1} + \text{BIS2} + \text{BIS3} + \text{BIS4} + (5 - \text{BIS5}) + \text{BIS6} + (5 - \text{BIS7})$$

If an item is missing, the total score should not be calculated. In this case, enter 88.

Higher scores are interpreted as reflecting higher levels of behavioral inhibition.

Form C5F: Interpersonal Reactivity Index¹ — INFORMANT QUESTIONNAIRE

The intent of the Interpersonal Reactivity Index (IRI) is to measure the subject's ability to empathize with others. Ask the informant to check the most accurate response for each item below. Tell the informant that "don't know" and "not applicable" are not allowable responses for any item. See *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* for more details on this questionnaire.

INSTRUCTIONS FOR THE CENTER

This questionnaire is designed to be completed **independently by the informant**, who will be describing the subject's current typical behavior. This form may be handed to the informant for completion by him- or herself at any time during the study visit. If the informant asks for clarification of questions, it is acceptable for a qualified psychologist or psychometrist to discuss the questionnaire with him or her. However, if the informant completes this questionnaire collaboratively with the clinician, either face-to-face or via telephone, **you must inform NACC of this change in protocol** by checking the appropriate box in the gray "FOR CLINIC USE ONLY" area at the top of the questionnaire.

Before the informant leaves, clinic staff should make sure that all questions were completed by the informant (i.e., none was left blank) by discussing the missing item with the informant and encouraging them to provide a response. If this is not done and it is later noticed that some items were missed by the informant, clinic staff should call the informant as soon as possible so that the missing items can be completed by phone. In this case, the questionnaire is not considered to have been completed independently by the informant. In the shaded area at the top of the form, the appropriate response would therefore be, "This questionnaire was completed via telephone interview of informant by clinic staff."

If there are still missing items, these items should be left blank, and "88" should be entered for the Empathic Concern (EC) Score and the Perspective-taking (PT) Score.

THIS QUESTIONNAIRE WAS COMPLETED:

- 0 Independently by co-participant, as described in "Instructions to the Center" 1 Via in-person interview of co-participant by clinic staff
 2 Via telephone interview of co-participant by clinic

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, select **3=Via telephone interview of co-participant by clinic staff**. Select only one method.

Please give us some information about yourself:

Your sex:

1 Male

2 Female

Your date of birth (mm/yyyy):

____ / ____ - ____

The co-participant should enter the month and year of his or her birth in the specified numerical format (e.g., March 1920 would be entered as “03/1920”). If the co-participant is unable or unwilling to answer, enter 99/9999. If only the year of birth is reported, enter “99” in the month field (e.g., 1920 would be entered as (“99/1920”).

Your relationship to subject:

1 Spouse or spouse equivalent

2 Child

3 Daughter- or son-in-law

4 Parent

5 Sibling

6 Other (other relative, friend, neighbor, paid caregiver)

DIRECTIONS: Indicate how well each statement describes the subject's **CURRENT** behavior. There are no right or wrong answers; we just want to get your impression of how you think the subject typically behaves.

If you have questions about how to complete this questionnaire, please ask a staff member, and they will be happy to help you.

Does NOT
describe
well



Describes
VERY
well

1. The subject shows tender, concerned feelings for people less fortunate than him/her.

1 2 3 4 5

2. The subject sometimes finds it difficult to see things from the “other guy’s” point of view.

1 2 3 4 5

3. Sometimes the subject does NOT feel very sorry for other people when they are having problems.

1 2 3 4 5

4. The subject tries to look at everybody’s side of a disagreement before he/she makes a decision.

1 2 3 4 5

5. If the subject sees somebody being taken advantage of, the subject feels kind of protective towards him/her.

1 2 3 4 5

6. The subject is likely to try to understand others better by imagining how things look from their perspective.

1 2 3 4 5

7. Other people’s misfortunes do NOT usually disturb the subject a great deal.

1 2 3 4 5

| | Does NOT describe well | ←-----→ | Describes VERY well | | |
|----------------------------------------------------------------------------------------------------------------------------------|------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 8. If the subject is sure he/she is right about something, he/she doesn't waste much time listening to other people's arguments. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 9. If the subject sees someone being treated unfairly, the subject doesn't feel much pity for him/her. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 10. The subject is often quite touched by things he/she sees happen. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 11. The subject believes that there are two sides to every question and tries to look at both of them. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 12. I would describe the subject as a pretty soft-hearted person. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 13. If the subject is upset at someone, the subject usually tries to put him/herself "in the other person's shoes" for a while. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 14. Before criticizing me, the subject is likely to imagine how he/she would feel if he/she were in my place. | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |

¹ Davis MH. *Measuring individual differences in empathy: evidence for a multidimensional approach*. J Pers Soc Psychol 1983; 44(1):113-126.

FOR CLINIC USE ONLY:

15. Empathic Concern Score (EC) (7–35): ___ ___

16. Perspective-taking Score (PT) (7–35): ___ ___

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SCORING INSTRUCTIONS FOR FORM C₅F

Each item yields a score from 1 to 5. Higher scores are interpreted as reflecting a greater degree of empathy.

The **Empathic Concern Score (EC)**, ranging from 7 to 35, is calculated by summing items 1, 3, 5, 7, 9, 10, and 12, as follows; note that the formula below performs the required *reverse scoring* of items 3, 7, and 9:

$$IRI1 + (6 - IRI3) + IRI5 + (6 - IRI7) + (6 - IRI9) + IRI10 + IRI12$$

If an item is missing, the total score should not be calculated. In this case, enter "88".

The **Perspective Taking Score (PT)**, ranging from 7 to 35, is calculated by summing items 2, 4, 6, 8, 11, 13, and 14, as follows; note that the formula below performs the required *reverse scoring* of items 2 and 8:

$$(6 - IRI2) + IRI4 + IRI6 + (6 - IRI8) + IRI11 + IRI13 + IRI14$$

If an item is missing, the total score should not be calculated. In this case, enter "88".

Form C6F: Revised Self-monitoring Scale¹ — INFORMANT QUESTIONNAIRE

The intent of the Revised Self-Monitoring Scale (RSMS) is to measure sensitivity to the expressive behavior of others and the ability to modify self-presentation. Ask the informant to check the most accurate response for each item below. Tell the informant that “don’t know” and “not applicable” are not allowable responses for any item. See *FTLD Module – Instructions for Neuropsychological Questionnaires (Forms C2F – C6F) and Tests Reported on Form C1F* for more details on this questionnaire.

INSTRUCTIONS FOR THE CENTER

This questionnaire is designed to be completed **independently by the informant**, who will be describing the subject’s current typical behavior. This form may be handed to the informant for completion by him- or herself at any time during the study visit. If the informant asks for clarification of questions, it is acceptable for a qualified psychologist or psychometrist to discuss the questionnaire with him or her. However, if the informant completes this questionnaire collaboratively with the clinician, either face-to-face or via telephone, **you must inform NACC of this change in protocol** by checking the appropriate box in the gray “FOR CLINIC USE ONLY” area at the top of the questionnaire.

Before the informant leaves, clinic staff should make sure that all questions were completed by the informant (i.e., none was left blank) by discussing the missing item with the informant and encouraging them to provide a response. If this is not done and it is later noticed that some items were missed by the informant, clinic staff should call the informant as soon as possible so that the missing items can be completed by phone. In this case, the questionnaire is not considered to have been completed independently by the informant. In the shaded area at the top of the form, the appropriate response would therefore be, “This questionnaire was completed via telephone interview of informant by clinic staff.”

If there are still missing items, these items should be left blank, and “88” should be entered for the Sensitivity to Socio-emotional Expressiveness (EX) Score, the Ability to Modify Self-presentation (SP) Score, and the RSMS Total Score.

THIS QUESTIONNAIRE WAS COMPLETED:

- 0 Independently by co-participant, as described in “Instructions to the Center” 1 Via in-person interview of co-participant by clinic staff
 2 Via telephone interview of co-participant by clinic

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, select **2=Via telephone interview of informant by clinic staff**. Select only one method.

DIRECTIONS: Indicate how well each statement describes the subject's **CURRENT** behavior. There are no right or wrong answers; we just want to get your impression of how you think the subject typically behaves. If you have questions about how to complete this questionnaire, please ask a staff member, and they will be happy to help you.

Certainly, always false (0) Generally false (1) Somewhat false, but with exceptions (2) Somewhat true, but with exceptions (3) Generally true (4) Certainly, always true (5)

| | | | | | | |
|----------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|----------------------------|----------------------------|----------------------------|
| 1. In social situations, the subject has the ability to alter his/her behavior if he/she feels that something else is called for. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 2. The subject is often able to correctly read people's true emotions through their eyes. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 3. The subject has the ability to control the way he/she comes across to people, depending on the impression he/she wants to give them. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 4. In conversations, the subject is sensitive to even the slightest change in the facial expression of the person he/she is conversing with. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 5. The subject's powers of intuition are quite good when it comes to understanding others. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 6. The subject can usually tell when others consider a joke in bad taste, even though they may laugh convincingly. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 7. When the subject feels that the image he/she is projecting isn't working, he/she can readily change to something that does. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 8. The subject can usually tell when he/she said something inappropriate by reading it in the listener's eyes. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 9. The subject has trouble changing his/her behavior to suit different people and different situations. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 10. The subject can adjust his/her behavior to meet the requirements of any situation he/she is in. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 11. If someone is lying to the subject, he/she usually knows it at once from that person's manner or expression. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 12. Even when it might be to his/her advantage, the subject has difficulty putting up a good front. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |
| 13. Once the subject knows what the situation calls for, it's easy for him/her to regulate his/her actions accordingly. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 2 | <input type="checkbox"/> 3 | <input type="checkbox"/> 4 | <input type="checkbox"/> 5 |

¹ Copyright © 1984 by the American Psychological Association. Adapted with permission. The official citation that should be used in referencing this material is Table 9 (adapted), p. 1361, from Revision of the Self-Monitoring Scale. Lennox, Richard D.; Wolfe, Raymond N. Journal of Personality and Social Psychology, Vol 46(6), Jun 1984, 1349-1364. doi: 10.1037/0022-3514.46.6.1349. No further reproduction or distribution is permitted without written permission from the American Psychological Association.

FOR CLINIC USE ONLY:

- | | |
|----------------------------------------------------------------------|---------|
| 14. Sensitivity to Socio-emotional Expressiveness Score (EX) (0–30): | ___ ___ |
| 15. Ability to Modify Self-presentation Score (SP) (0–35): | ___ ___ |
| 16. RSMS Total Score (0–65): | ___ ___ |

SCORING INSTRUCTIONS FOR FORM C6F

Each item yields a score from 0 to 5. Higher scores are interpreted as reflecting a greater degree of interpersonal sensitivity and responsiveness.

- The **Sensitivity to Socioemotional Expressiveness Score (EX)**, ranging from 0 to 30, is the sum of items 2, 4, 5, 6, 8, and 11. The EX score may be calculated with the following formula:

$$\text{RSMS2} + \text{RSMS4} + \text{RSMS5} + \text{RSMS6} + \text{RSMS8} + \text{RSMS11}$$

If an item is missing, the total score should not be calculated. In this case, enter “88”.

- The **Ability to Modify Self-Presentation Score (SP)**, ranging from 0 to 35, is calculated by summing items 1, 3, 7, 9, 10, 12, and 13, as follows; note that the formula below performs the required reverse scoring of items 9 and 12:

$$\text{RSMS1} + \text{RSMS3} + \text{RSMS7} + (5 - \text{RSMS9}) + \text{RSMS10} + (5 - \text{RSMS12}) + \text{RSMS13}$$

If an item is missing, the total score should not be calculated. In this case, enter “88”.

- The **RSMS Total Score**, ranging from 0 to 65, is the sum of all 13 items, as follows; note that the formula below performs the required reverse scoring of items 9 and 12:

$$\text{RSMS1} + \text{RSMS2} + \text{RSMS3} + \text{RSMS4} + \text{RSMS5} + \text{RSMS6} + \text{RSMS7} + \text{RSMS8} + (5 - \text{RSMS9}) + \text{RSMS10} + \text{RSMS11} + (5 - \text{RSMS12}) + \text{RSMS13}$$

If an item is missing, the total score should not be calculated. In this case, enter “88”.

Form E2F: Imaging Available

The purpose of this form is to record whether imaging is available from the subject's current visit or previous visits. The form should be completed by the principal clinician or imaging specialist involved in interpreting the scan(s).

INSTRUCTIONS: This form is to be completed by the clinician or imaging specialist involved in interpreting the scan. For additional clarification and examples, see FTLD Module Coding Guidebook for Follow-up Visit Packet, Form E2F. Check only one box per question.

- 1. Is a structural MRI scan, obtained as part of the current evaluation or a previous evaluation and not yet recorded, available for data sharing? (REPORT MOST RECENT)**
- 0 No
 1 Yes

IF YES, complete 1a – 1f; if no, go to Question 2.

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

___ / ___ / _____

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. Is it in DICOM format or other electronic format?

- 0 No
 1 Yes (specify format):

 9 Unknown

- 1c. Was ADNI protocol used?

- 0 No
 1 Yes
 ADNI version: _____
 9 Unknown

For information on Alzheimer's Disease Neuroimaging Initiative (ADNI) protocol, see Jack CR Jr, Bernstein MA, Fox NC, et al. The Alzheimer's Disease Neuroimaging Initiative (ADNI): MRI methods. *J Magn Reson Imaging* 2008; 27(4):685-691.

- 1d. Scan manufacturer:

- 1 GE
 2 Siemens
 3 Philips
 4 Other: _____
 9 Unknown

| | |
|----------------------------------------------|-------|
| 1d1. Scan model: | _____ |
| If scan model is not known, enter “Unknown”. | |

| | |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1e. Field strength: | <input type="checkbox"/> 1 1.5T <input type="checkbox"/> 2 3T <input type="checkbox"/> 3 7T <input type="checkbox"/> 4 Other: _____ <input type="checkbox"/> 9 Unknown |
| 1f. Are results of quantitative image analysis available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| 2. Is an FDG-PET scan, obtained as part of the current evaluation or a previous evaluation and not yet recorded, available for data sharing? (REPORT MOST RECENT) | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| IF YES, complete 2a – 2e; if no, go to Question 3. | |

| | |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------|
| 2a. Date of scan (MM / DD / YYYY): <i>NOTE: A value of 99 (unknown) is permissible for day only.</i> | ____ / ____ / _____ |
| 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. Is it in DICOM format or other electronic format? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes (specify format): _____ <input type="checkbox"/> 9 Unknown |
|-------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------|

| | |
|-----------------------------|-------------------------------------------------------------------------------------------------------------------------------------|
| 2c. Was ADNI protocol used? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <i>ADNI version:</i> _____ <input type="checkbox"/> 9 Unknown |
|-----------------------------|-------------------------------------------------------------------------------------------------------------------------------------|

For information on Alzheimer’s Disease Neuroimaging Initiative (ADNI) protocol, see Jack CR Jr, Bernstein MA, Fox NC, et al. The Alzheimer’s Disease Neuroimaging Initiative (ADNI): MRI methods. *J Magn Reson Imaging* 2008; 27(4):685-691.

| | |
|------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 2d. Scan manufacturer: | <input type="checkbox"/> 1 GE <input type="checkbox"/> 2 Siemens <input type="checkbox"/> 3 Philips <input type="checkbox"/> 4 Other: _____ <input type="checkbox"/> 9 Unknown |
|------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|

| | |
|----------------------------------------------|-------|
| 2d1. Scan model: | _____ |
| If scan model is not known, enter "Unknown". | |

| | |
|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------|
| 2e. Are results of quantitative image analysis available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| 3. Is an amyloid PET scan, obtained as part of the current evaluation or a previous evaluation and not yet recorded, available for data sharing? (REPORT MOST RECENT) | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| IF YES, complete 3a – 3f; if no, go to Question 4. | |

| | |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------|
| 3a. Date of scan (MM / DD / YYYY): <i>NOTE: A value of 99 (unknown) is permissible for day only.</i> | ____ / ____ / _____ |
| 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. Is it in DICOM format or other electronic format? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes (specify format): _____ <input type="checkbox"/> 9 Unknown |
| 3c. Ligand used: | <input type="checkbox"/> 1 11C-PIB <input type="checkbox"/> 2 18F-AV45 <input type="checkbox"/> 3 Other (specify): _____ <input type="checkbox"/> 9 Unknown |

| | |
|-----------------------------|-------------------------------------------------------------------------------------------------------------------------------------|
| 3d. Was ADNI protocol used? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <i>ADNI version:</i> _____ <input type="checkbox"/> 9 Unknown |
|-----------------------------|-------------------------------------------------------------------------------------------------------------------------------------|

For information on Alzheimer’s Disease Neuroimaging Initiative (ADNI) protocol, see Jack CR Jr, Bernstein MA, Fox NC, et al. The Alzheimer’s Disease Neuroimaging Initiative (ADNI): MRI methods. *J Magn Reson Imaging* 2008; 27(4):685-691.

| | |
|------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 3e. Scan manufacturer: | <input type="checkbox"/> 1 GE <input type="checkbox"/> 2 Siemens <input type="checkbox"/> 3 Philips <input type="checkbox"/> 4 Other: _____ <input type="checkbox"/> 9 Unknown |
|------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|

| | |
|------------------|-------|
| 3e1. Scan model: | _____ |
|------------------|-------|

If scan model is not known, enter “Unknown”.

| | |
|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------|
| 3f. Are results of quantitative image analysis available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| 4. Are other PET or SPECT scans, obtained as part of the current evaluation or a previous evaluation and not yet recorded, available for data sharing? (REPORT MOST RECENT) | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes |
| IF YES, complete 4a – 4d; if no, END FORM HERE. | |
| 4a. Is a dopaminergic scan available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown |
| 4b. Is a serotonergic scan available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown |
| 4c. Is a cholinergic scan available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown |
| 4d. Is another kind of scan available? | <input type="checkbox"/> 0 No <input type="checkbox"/> 1 Yes (SPECIFY): _____ |

Form E3F: Imaging in Diagnosis

The purpose of this form is to record whether imaging is available from the subject's current visit. The form should be completed by the clinician or imaging specialist involved in interpreting the scan(s).

INSTRUCTIONS: This form is to be completed by the clinician or imaging specialist involved in interpreting the scan. For additional clarification and examples, see FTLD Module Coding Guidebook for Follow-up Visit Packet, Form E3F. Check only one box per question.

| | No | Yes | Unknown |
|--------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|---------|
| 1. Was imaging obtained as part of this visit for use in diagnosis? If the answer is "0 (No)", SKIP THE REST OF THIS FORM. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |

STRUCTURAL MRI

| | | | |
|--------------------------------------------------------------------|----------------------------|----------------------------|--|
| 2. Was structural MRI done? If "No", SKIP TO QUESTION 3. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |
|--------------------------------------------------------------------|----------------------------|----------------------------|--|

If structural MRI was not done, skip this section.

| | | | |
|-----------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 2a. Was focal atrophy (beyond what would be expected for age) appreciated by visual inspection? If "No" or "Unknown", SKIP TO QUESTION 3. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
|-----------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|

Select **9=Unknown** only if focal atrophy status is unknown.

Questions 2a1 – 2a11: Select **9=Unknown** only if focal atrophy status is unknown in the specified location.

| Where was focal atrophy appreciated? | No | Yes | Unknown |
|--------------------------------------|----------------------------|----------------------------|----------------------------|
| 2a1. Right frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a2. Left frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a3. Right temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a4. Left temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a5. Right medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a6. Left medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a7. Right parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a8. Left parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

| | No | Yes | Unknown |
|---------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 2a9. Right basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a10. Left basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 2a11. Other area of the brain (specify below): _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

FDG-PET

3. Was FDG-PET done?
If "No", SKIP TO QUESTION 4.

0 1

If FDG-PET was not done, skip this section.

3a. Was focal hypometabolism appreciated by visual inspection?
If "No" or "Unknown", SKIP TO QUESTION 4.

0 1 9

Select **9=Unknown** only if focal hypometabolism status is unknown.

Questions 3a1 – 3a11: Select **9=Unknown** only if focal hypometabolism status is unknown in the specified location.

Where was focal hypometabolism appreciated?

| | | | |
|---------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 3a1. Right frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a2. Left frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a3. Right temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a4. Left temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a5. Right medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a6. Left medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a7. Right parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a8. Left parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a9. Right basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a10. Left basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 3a11. Other area of the brain (specify below): _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

| | No | Yes | Unknown |
|-----------------------------------------------------------------|----------------------------|----------------------------|---------|
| AMYLOID PET | | | |
| 4. Was amyloid PET done? If "No", SKIP TO QUESTION 5. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |

If FDG-PET was not done, skip this section.

| | | | |
|------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 4a. Was amyloid deposition appreciated by visual inspection? If "No" or "Unknown", SKIP TO QUESTION 5. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
|------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|

Select **9=Unknown** only if amyloid deposition status is unknown.

Questions 4a1 – 4a11: Select **9=Unknown** only if amyloid deposition status is unknown in the specified location.

| Where was amyloid deposition noted? | | | |
|---------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 4a1. Right frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a2. Left frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a3. Right temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a4. Left temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a5. Right medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a6. Left medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a7. Right parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a8. Left parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a9. Right basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a10. Left basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 4a11. Other area of the brain (specify below): _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

| | No | Yes | Unknown |
|---------------------------------------------------------------|----------------------------|----------------------------|---------|
| CBF SPECT | | | |
| 5. Was CBF SPECT done? If "No", SKIP TO QUESTION 6. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |

If CBF SPECT was not done, skip this section.

| | No | Yes | Unknown |
|---------------------------------------------------------------------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 5a. Were abnormalities appreciated by visual inspection? If "No" or "Unknown", SKIP TO QUESTION 6. | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

Select **9=Unknown** only if the status of CBF SPECT-detected abnormalities is unknown.

Questions 5a1 – 5a11: Select **9=Unknown** only if the status of CBF SPECT-detected abnormalities is unknown in the specified location.

| Where were abnormalities noted? | | | |
|---------------------------------------------------------|----------------------------|----------------------------|----------------------------|
| 5a1. Right frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a2. Left frontal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a3. Right temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a4. Left temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a5. Right medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a6. Left medial temporal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a7. Right parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a8. Left parietal lobe | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a9. Right basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a10. Left basal ganglia | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |
| 5a11. Other area of the brain (specify below): _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | <input type="checkbox"/> 9 |

| | No | Yes | Unknown |
|----------------------------------------------------------------------|----------------------------|----------------------------|---------|
| OTHER IMAGING | | | |
| 6. Was other imaging done? If "Yes", specify: _____ | <input type="checkbox"/> 0 | <input type="checkbox"/> 1 | |