NACC Uniform Data Set (UDS) — FTLD Module

Coding Guidebook

FOR FOLLOW-UP VISIT PACKET

Detailed, annotated explanations of each form on an item-level basis, with instructions, operational definitions, and references

NOTE: Version 2 is NOT the most current version of the FTLD Moudle forms and is no longer used for data submission. For the most current version, please visit http://www.alz.washington.edu.

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NACC
National Alzheimer's Coordinating Center

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 a light gray background or are contained in a light gray box. To make it easier to locate the Guidebook material, all gray shading has been removed from the reproductions of the forms themselves.

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 annual visits following the initial enrollment into the UDS 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.** 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

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

NACC expects and intends that all FTLD forms will 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 informant, 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 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 indicate 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 information

Form	Description	Submitted: Yes	Submitted: No	If not submitted, specify reason (see key, above)	Comments (provide if needed)
A3aF	Record of Consent for Biologic Specimen Use	☐ 1	☐ 0	____ ____	
A3F	Family History: Affected Family Members	☐ 1	☐ 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	☐ 1	☐ 0	____ ____	
C5F	Interpersonal Reactivity Index	☐ 1	☐ 0	____ ____	
C6F	Revised Self-monitoring Scale	☐ 1	☐ 0	____ ____	
E2F	Imaging Available	**Required**			
E3F	Imaging in Diagnosis	**Required**			

Check "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, check "No" and enter the appropriate Key code for the reason and provide a written explanation in the "Comments" section.

Form A3aF: Record of Consent for Biologic Specimen Use

NOTE: This form is to be completed by clinic staff responsible for obtaining consents, based on an existing consent at clinic. For additional clarification and examples, 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. If the information below was previously submitted for the relative but there has been a change in consent, fill out the form in its entirety. If information recorded previously is found to be incorrect (e.g. sibling or child's birth date), edit the original form; do not create a new entry on this form.

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 and FTLD Module Initial Visit or FTLD Follow-up Visit Form A3F (if applicable).

☐ 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:

a. Uniform Data Set (UDS) Initial Visit Form A3 or UDS Follow-up Visit Form A3,
b. FTLD Module Initial Visit Form A3aF or FTLD Module Follow-up Visit Form A3aF (if applicable), and
c. FTLD Module Initial Visit Form A3F or FTLD Module Follow-up Visit Form A3F (if applicable).

An FTLD Module FVP Form A3aF is necessary under the following circumstances:

1. **New consent** — If any relatives not previously recorded have newly provided consent, fill this form out in its entirety for each new consent.
2. **Change in consent** — If a relative withdraws consent entirely, fill out a new FTLD FVP Form A3aF and select 'No' for Questions 1a through 1c. A new Form A3aF would also be submitted if there were a change in consent for one or more items 1a through 1c. For example, if the subject's father previously said that he did not want his sample to be given to researchers at other institutions, but he has since changed his mind, fill out and submit a new FTLD FVP Form A3aF for the subject's father.

An FTLD Module FVP Form A3aF form is NOT necessary under the following circumstances:

1. **No change in consent and no new consents provided.**
2. **Erroneous information** (a mistake is found in previously provided information for Form A3aF) — For example, if a sibling who previously provided consent was recorded as being born in 1938, when the subject's sibling providing consent was actually born in 1939, edit the original form; do not create a new entry on this form. If the subject's mother was thought to have provided consent, when in reality it was the subject's father, edit the original form; do not create a new entry on this form.

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).	☐ 0 No	☐ 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.	☐ 0 No	☐ 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 would happen, I would not be offered a share in any profits. I permit (home institution) to give my sample to researchers at other institutions.	☐ 0 No	☐ 1 Yes

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

Form A3F: Family History: Affected Family Members

NOTE: This form is to be completed by a clinician with experience in evaluating patients with frontotemporal lobar degeneration. For additional clarification and examples, see FTLD Coding Guidebook for Follow-up Visit Packet, Form A3F.

INSTRUCTIONS: Review all data collected on A3F at the previous FTLD module visit(s), if applicable, with the subject or informant. For family members who were indicated as being affected at a previous FTLD module visit, any corrections to their data should be made to the previous A3F form. Any newly obtained diagnoses for a family member, including family members previously reported as being affected at a past FTLD module visit, should be indicated on this form and should not be submitted as a correction to the previous A3F.

If this is the first time you are providing A3F information for this subject's family, fill out this form in its entirety.

"AFFECTED FAMILY MEMBERS" — Please consider blood relatives only. For the purposes of Form A3F, "affected" means affected by dementia **OR** by any of the non-normal clinical diagnoses listed in Appendix 1 on page 6 of this form.

If it was discovered that the family has a new predominant mutation, that supersedes a mutation reported on a previous A3F, then include this new information on this form. However, if the wrong mutation was indicated on a previous A3F, make the correction to that previous form. Any new information on a family member, obtained since the last FTLD module visit, should be reported on this form.

1. AFFECTED FAMILY MEMBERS			
Since the last FTLD Module visit, is new information available concerning data collected by items 1a through 1g, below? **If 1 (Yes), COMPLETE SECTION 1 and then go on to Section 2.** **If 0 (No), SKIP TO SECTION 2.**	☐ 0 No	☐ 1 Yes	
1a. Are there affected family members? (See box above for definition of "affected.") **If the answer is "No" or "Unknown," please skip the rest of this form.**	☐ 0 No	☐ 1 Yes	☐ 9 Unknown

Only family members who are affected by dementia or have a non-normal clinical diagnosis should be listed on this form.

1b. In this family, is there a known mutation in a gene associated with FTLD? **If the answer is "No" or "Unknown," please SKIP TO SECTION 2.**	☐ 0 No	☐ 1 Yes	☐ 9 Unknown

Indicate whether blood relatives have a genetic mutation associated with FTLD.

1c. What is the predominant mutation?	☐ 1 MAPT ☐ 2 PGRN ☐ 3 C9ORF72 ☐ 4 FUS ☐ 8 Other (SPECIFY: _____) ☐ 9 Unknown		
Although blood relatives might have more than one known genetic mutation, indicate the predominant one.			
1d. Is there evidence for this mutation in the form of commercial lab test documentation?	☐ 0 No	☐ 1 Yes	☐ 9 Unknown
1e. Is there evidence for this mutation in the form of research lab test documentation?	☐ 0 No	☐ 1 Yes	☐ 9 Unknown
1f. Is there evidence for this mutation in the form of family report?	☐ 0 No	☐ 1 Yes	☐ 9 Unknown
1g. Is there other evidence for this mutation?	☐ 0 No ☐ 1 Yes (SPECIFY: _____) ☐ 9 Unknown		

AFFECTED PARENTS — Use the form below to provide information on affected parents only (see definition of "affected" in the box above). **Provide all information below if it has not been submitted previously. If you are updating previously submitted A3F data for one or more relatives, please enter all data in the row for that relative and also correct any previously submitted A3F form data for this relative, if applicable. Otherwise, check the box for 0 (No) in the first line below to indicate no affected parent or no change since data were previously submitted on affected parents.**

2. AFFECTED PARENTS				
Since the last FTLD Module visit, is new information available concerning the status of the subject's mother or father? **If 1 (Yes), please COMPLETE ITEM 2a and/or ITEM 2b, below. If 0 (No), SKIP SECTION 2 and go on to Section 3.**			☐ 0 No	☐ 1 Yes
	a. Neurological problem*	**b. Primary DX****	**c. Method of evaluation*****	**d. Age of onset**
2a. Mother	___	___ ___ ___	___	___ ___ ___
2b. Father	___	___ ___ ___	___	___ ___ ___

If previously reported information is found to have been reported in error (e.g., a mistaken age of onset or an incorrectly coded neurological problem), edit the original form.

Following are scenarios with examples of "new information" about parents:

1. **New diagnosis** — A subject's parent is newly diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, or the new informant has information on the parent's dementia that the previous informant did not. Example: At the time of the FTLD Module Initial Visit, the subject's mother had complained about her memory but had not had a clinical evaluation. No information on the subject's mother was recorded on FTLD IVP Form A3F. Since then, the subject's mother saw a clinician and was diagnosed with mild cognitive impairment. Check Yes (1) in the first row and fill out the appropriate information on line 2a. *Do not change the original form.*
2. **Change in diagnosis** — A subject's parent was diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, but the diagnosis has changed based on clinician judgment (e.g., due to decline, improvement, or change in symptoms). Example: The subject's father was diagnosed with Alzheimer's disease two years ago, and these data were recorded on the subject's FTLD Module IVP Form A3F. Since then, the subject's father has had an examination in which he received a primary diagnosis of behavioral variant FTD. Check Yes (1) in the first row and fill out the appropriate new information on line 2b. *Do not correct the original form.* Age of onset should reflect the age at which symptoms began, not the age at which this new diagnosis was made.
3. **Change in method of evaluation** — A subject's parent has a new evaluation method for an existing diagnosis or neurological problem. Example: The subject's father was diagnosed with Parkinson's disease (PD), and "Examination" (2) was entered for the Method of Evaluation on the subject's FTLD Module IVP Form A3F. The subject's father has since died and undergone autopsy and now has an autopsy-confirmed PD diagnosis. Check Yes (1) in the first row and fill out the appropriate new information on line 3a. *Do not correct the original form.*

When entering a code for the primary diagnosis ("Primary DX"), select the most specific diagnosis that is known.

If the method of evaluation is unknown, select "Family report." If multiple methods were used, report the highest level of diagnostic evaluation (see Appendix 2 for an explanation of the methods of evaluation and their ranking from highest [1] to lowest [7]).

Age of onset refers to the age at which symptoms began, not the age at which diagnosis was made. Symptoms used to determine age of onset should reflect significant functional change in the individual's abilities (e.g., judgment, orientation, management of personal finances, and conduct of home activities). If the subject or informant is unwilling or unable to answer, enter 999. If the primary diagnosis is a congenital condition, or present since birth, indicate an age of onset of 0.

***Codes for neurological problems and psychiatric conditions**

1 Cognitive impairment/ behavior change
2 Parkinsonism
3 ALS
4 Other neurologic condition such as multiple sclerosis or stroke
5 Psychiatric condition such as schizophrenia, bipolar disorder, or alcoholism
9 Unknown

****Codes for primary diagnosis**

See Appendix 1 on page 6 of this form

*****Codes for method of evaluation**

For descriptions, see Appendix 2 on page 6 of this form

1 Autopsy
2 Examination
3 Medical record review from formal dementia evaluation
4 Review of general medical records AND informant and/or subject telephone interview
5 Review of general medical records only
6 Subject and/or informant telephone interview
7 Family report

AFFECTED SIBLINGS — Use the form below to provide information on affected siblings only (see definition of "affected" in the box on page 1 of this form). **Provide all information below if it has not been submitted previously. If you are updating previously submitted A3F data for one or more relatives, please enter all data in the row for that relative and also correct any previously submitted A3F form data for this relative, if applicable. Otherwise, check the box for 0 (No) in the first line below to indicate no affected sibling or no change since data were previously submitted on affected siblings.**

"Sibling's birth year" on this form MUST agree with the birth year listed for that sibling on UDS Initial Visit or UDS Follow-up Visit Form A3 and FTLD Module Initial Visit or FTLD Follow-up Visit Form A3F (if applicable).

"Unknown" (9999) is not a permissible value. If birth year is unknown, please provide an approximate year on UDS Initial Visit Form A3 so that the sibling with unknown birth year ends up in correct birth order relative to the other siblings. (EXAMPLE: Suppose a subject is the oldest of three children. The subject was born in 1930 and the middle sibling in 1933; the youngest sibling's birth year is unknown. An approximate birth year of 1934 or later should be assigned to the youngest sibling.) **Use that same birth year on FTLD Module Forms A3F and A3aF.**

If an affected sibling has already been listed on UDS Initial Visit Form A3 with a birth year of 9999, then UDS Initial Visit Form A3 must be edited so that an approximate birth year is entered, as described in the paragraph above. That same birth year should be entered below.

"Sibling's birth month" should be filled out if known; otherwise, please enter "99". Only full siblings should be listed.

3. AFFECTED SIBLINGS

Since the last FTLD Module visit, is new information available concerning the status of any of the subject's siblings? **If 1 (Yes), please COMPLETE SECTION 3, below. If 0 (No), SKIP SECTION 3 and go on to Section 4.**	☐ 0 No	☐ 1 Yes

	a. Sibling's birth mo / yr	b. Neurological problem*	c. Primary DX**	d. Method of evaluation***	e. Age of onset
3a.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3b.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3c.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3d.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3e.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3f.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3g.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3h.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3i.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3j.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3k.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3l.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___
3m.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___

***Codes for neurological problems and psychiatric conditions**

1 Cognitive impairment/ behavior change
2 Parkinsonism
3 ALS
4 Other neurologic condition such as multiple sclerosis or stroke
5 Psychiatric condition such as schizophrenia, bipolar disorder, or alcoholism
9 Unknown

****Codes for primary diagnosis**

See Appendix 1 on page 6 of this form

*****Codes for method of evaluation**

For descriptions, see Appendix 2 on page 4 of this form

1 Autopsy
2 Examination
3 Medical record review from formal dementia evaluation
4 Review of general medical records AND informant and/or subject telephone interview
5 Review of general medical records only
6 Subject and/or informant telephone interview
7 Family report

If previously reported information is found to have been reported in error (e.g., a mistaken birth year, or an incorrectly coded neurological problem), edit the original form.

Following are scenarios with examples of "new information" about siblings:

1. **New diagnosis** — A subject's sibling is newly diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, or the new informant has information on the parent's dementia that the previous informant did not. Example: At the time of the FTLD Module Initial Visit, the subject's brother had complained about his memory but had not had a clinical evaluation. No information on the subject's brother was recorded on FTLD IVP Form A3F. Since then, the subject's brother saw a clinician and was diagnosed with mild cognitive impairment. Check Yes (1) in the first row and fill out the appropriate information on line 3a.
2. **Change in diagnosis** — A subject's sibling was diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, but the diagnosis has changed based on clinician judgment (e.g., due to decline, improvement, or change in symptoms). Example: The subject's brother was diagnosed with Alzheimer's disease two years ago, and these data were recorded on the subject's FTLD Module IVP Form A3F. Since then, the subject's brother had an examination in which he received a primary diagnosis of behavioral variant FTD. Check Yes (1) in the first row and fill out the appropriate new information on line 3a. *Do not correct the original form.* Age of onset should reflect the age at which symptoms began, not the age at which this new diagnosis was made.
3. **Change in method of evaluation** — A subject's sibling has been evaluated by a new method for an existing diagnosis or neurological problem. Example: The subject's brother was diagnosed with Parkinson's disease (PD), and "Examination" (2) was entered for the Method of Evaluation on the subject's FTLD Module IVP Form A3F. The subject's brother has since died and undergone autopsy and now has an autopsy-confirmed PD diagnosis. Check Yes (1) in the first row and fill out the appropriate new information on line 3a. *Do not correct the original form.*

Birth year for a given sibling should be constant across forms:

a. Uniform Data Set (UDS) Initial Visit Form A3 or UDS Follow-up visit Form A3,
b. FTLD Module Initial Visit Form A3aF or FTLD Module Follow-up Visit Form A3aF (if applicable), and
c. FTLD Module Initial Visit Form A3F or FTLD Module Follow-up Visit Form A3F (if applicable).

When entering a code for the primary diagnosis ("Primary DX"), select the most specific diagnosis that is known.

If the method of evaluation is unknown, select "Family report." If multiple methods were used, report the highest level of diagnostic evaluation (see Appendix 2 for an explanation of the methods of evaluation and their ranking from highest [1] to lowest [7]).

Age of onset refers to the age at which symptoms began, not the age at which diagnosis was made. Symptoms used to determine age of onset should reflect significant functional change in the individual's abilities (e.g., judgment, orientation, management of personal finances, and conduct of home activities). If the subject or informant is unwilling or unable to answer, enter 999. If the primary diagnosis is a congenital condition, or present since birth, indicate an age of onset of 0.

AFFECTED CHILDREN — Use the form below to provide information on affected children only (see definition of "affected" in the box on page 1 of this form). **Provide all information below if it has not been submitted previously. If you are updating previously submitted A3F data for one or more relatives, please enter all data in the row for that relative and also correct any previously submitted A3F form data for this relative, if applicable. Otherwise, check the box for 0 (No) in the first line below to indicate no affected child or no change since data were previously submitted on affected children.**

"Child's birth year" on this form MUST agree with the birth year listed for that sibling on UDS Initial Visit or UDS Follow-up Visit Form A3 and FTLD Module Initial Visit or FTLD Follow-up Visit Form A3F (if applicable)**.**

"Unknown" (9999) is not a permissible value. If birth year is unknown, please provide an approximate year on UDS Initial Visit Form A3 so that the child with unknown birth year ends up in correct birth order relative to the other children. (EXAMPLE: Suppose a subject has three children. The oldest is a son born in 1960, the youngest a son born in 1964, and the middle child a girl whose birth year is unknown. The girl should be assigned an approximate birth year of 1962 or 1963.) **Use that same birth year from UDS Initial Visit Form A3 on FTLD Module Forms A3F and A3aF.**

If an affected child has already been listed on UDS Initial Visit Form A3 with a birth year of 9999, then UDS Initial Visit Form A3 must be edited so that an approximate birth year is entered, as described in the paragraph above. That same birth year should be entered below.

"Child's birth month" should be filled out if known; otherwise, please enter "99".

4. AFFECTED CHILDREN

Since the last FTLD Module visit, is new information available concerning the status of any of the subject's children? **If 1 (Yes), please COMPLETE SECTION 4, below. If 0 (No), then END FORM HERE.**	☐ 0 No	☐ 1 Yes

	a. Child's birth mo / yr	b. Neurological problem*	c. Primary DX**	d. Method of evaluation***	e. Age of onset
4a.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4b.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4c.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4d.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4e.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4f.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4g.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4h.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4i.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4j.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4k.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4l.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __
4m.	__ __ / __ __ __ __	__	__ __ __	__	__ __ __

***Codes for neurological problems and psychiatric conditions**

1 Cognitive impairment/ behavior change
2 Parkinsonism
3 ALS
4 Other neurologic condition such as multiple sclerosis or stroke
5 Psychiatric condition such as schizophrenia, bipolar disorder, or alcoholism
9 Unknown

****Codes for primary diagnosis**

See Appendix 1 on page 6 of this form

*****Codes for method of evaluation**

For descriptions, see Appendix 2 on page 4 of this form

1 Autopsy
2 Examination
3 Medical record review from formal dementia evaluation
4 Review of general medical records AND informant and/or subject telephone interview
5 Review of general medical records only
6 Subject and/or informant telephone interview
7 Family report

If previously reported information is found to have been reported in error (e.g., a mistaken birth year, or an incorrectly coded neurological problem), edit the original form.

Following are scenarios with examples of "new information" about children:

1. **New diagnosis** — A subject's child is newly diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, or the new informant has information on the parent's dementia that the previous informant did not. For example, at the time of the FTLD Module Initial Visit, the subject's son had some mild behavioral symptoms but had not had a clinical evaluation. No information on the subject's son was recorded on the FTLD IVP Form A3F. Since then, the subject's son saw a clinician and was diagnosed with behavioral variant FTD. Check Yes (1) in the first row and fill out the appropriate information on line 4a.
2. **Change in diagnosis** — A subject's child was diagnosed with dementia or one of the non-normal clinical diagnoses listed in Appendix 1, but the diagnosis has changed based on clinician judgment (e.g., due to decline, improvement, or change in symptoms). For example, the subject's daughter was diagnosed with mild cognitive impairment two years ago, and these data were recorded on the subject's FTLD Module IVP Form A3F. Since then, the subject's daughter had an examination in which she received a primary diagnosis of probable Alzheimer's disease. Check Yes (1) in the first row and fill out the appropriate new information on line 3a. *Do not correct the original form.* Age of onset should reflect the age at which symptoms began, not the age at which this new diagnosis was made.
3. **Change in method of evaluation —** A subject's child has been evaluated by a new method for an existing diagnosis or neurological problem. Example: The subject's son was diagnosed with multiple sclerosis, and "Examination" (2) was entered for the Method of Evaluation on the subject's FTLD Module IVP Form A3F. The subject's son has since died and undergone autopsy and now his MS diagnosis is confirmed by autopsy. Check Yes (1) in the first row and fill out the appropriate new information on line 3a. *Do not correct the original form.*

Birth year for a given child should be constant across forms:

a. Uniform Data Set (UDS) Initial Visit Form A3 or UDS Follow-up visit Form A3,
b. FTLD Module Initial Visit Form A3aF or FTLD Module Follow-up Visit Form A3aF (if applicable), and
c. FTLD Module Initial Visit Form A3F or FTLD Module Follow-up Visit Form A3F (if applicable).

When entering a code for the primary diagnosis ("Primary DX"), select the most specific diagnosis that is known.

If the method of evaluation is unknown, select "Family report." If multiple methods were used, report the highest level of diagnostic evaluation (see Appendix 2 for an explanation of the methods of evaluation and their ranking from highest [1] to lowest [7]).

Age of onset refers to the age at which symptoms began, not the age at which diagnosis was made. Symptoms used to determine age of onset should reflect significant functional change in the individual's abilities (e.g., judgment, orientation, management of personal finances, and conduct of home activities). If the subject or informant is unwilling or unable to answer, enter 999. If the primary diagnosis is a congenital condition, or present since birth, indicate an age of onset of 0.

**APPENDIX 1: PRIMARY DIAGNOSIS CODES

CODE	DIAGNOSIS
040	Mild cognitive impairment (MCI), not otherwise specified
041	MCI — amnestic
042	MCI — multiple domain with amnesia
043	MCI — single domain nonamnestic
044	MCI — multiple domain nonamnestic
045	Impaired, but not MCI
050	Alzheimer's disease
070	Dementia with Lewy bodies
080	Vascular dementia
100	Alcohol-related dementia
110	Dementia of undetermined etiology
120	Behavioral variant frontotemporal dementia
130	Primary progressive aphasia, semantic variant
131	Primary progressive aphasia, nonfluent/agrammatic variant
132	Primary progressive aphasia, logopenic variant
133	Primary progressive aphasia, not otherwise specified

CODE	DIAGNOSIS
140	Progressive supranuclear palsy
150	Corticobasal syndrome/corticobasal degeneration
160	Huntington's disease
170	Prion disease
180	Cognitive dysfunction from medications
190	Cognitive dysfunction from medical illness
200	Depression
210	Other major psychiatric illness
220	Down syndrome
230	Parkinson disease
240	Stroke
250	Hydrocephalus
260	Traumatic brain injury
270	CNS neoplasm
280	Other
310	Amyotrophic lateral sclerosis
320	Multiple sclerosis

If the method of evaluation is by subject and/or informant telephone interview or by family report and the primary diagnosis is unknown, enter 999 for primary diagnosis. If the neurological problem and the primary diagnosis are unknown, leave the entire row blank.

***APPENDIX 2: METHOD OF EVALUATION

1. **Autopsy —** If the autopsy was performed at an outside institution, **you must have the report** to code as diagnosis by autopsy.
2. **Examination —** The subject must have been examined in person at your ADC/institution or by genetic studies staff associated with your ADC/institution to code as diagnosis by examination. Medical records may or may not have been used when assigning diagnosis.
3. **Medical record review from formal dementia evaluation —** Medical records should be from an examination that focused specifically on dementia; that was performed by a neurologist, geriatrician, or psychiatrist; that includes a neurologic examination, an imaging study, and cognitive testing (e.g., MMSE, Blessed, or more formal tests). A telephone interview may also be used to collect additional information.
4. **Review of general medical records AND informant and/or subject telephone interview — General medical records** can be of various types, including those from a primary-care physician's office, hospitalization records, nursing home records, etc. They may include a neurologic exam and a cognitive test such as the MMSE along with a medical history. **The telephone interview** with the subject and/or the informant should include a medical history to capture the nature and presentation of cognitive deficits, if present, and age of onset if symptomatic. If the subject is normal or is in the early stages of dementia, brief formal cognitive testing should be included in the interview. Unless an affected subject is in the early stages of dementia, the interview should be conducted with an informant.
5. **Review of general medical records ONLY —** See definition No. 4 above. If general medical records are used to diagnose a subject as demented or not demented, they should include a medical history, neurologic exam, and a cognitive test such as MMSE. In most cases, general medical records alone should not be used to assign a diagnosis of mild cognitive impairment, or of any of the FTLD spectrum subtypes, or of parkinsonian disorders other than Parkinson disease.
6. **Subject and/or informant telephone interview —** See definition No. 4 above.
7. **Family report —** Family report should be coded when the informant for the family reports a subject as having been diagnosed with a particular disorder. In most cases, family report alone should not be used to assign a diagnosis of mild cognitive impairment, or of any of the FTLD spectrum subtypes, or of parkinsonian disorders other than Parkinson disease.

Form B3F: Supplemental UPDRS

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

NOTE: 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.

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

		Not to a degree that would justify such a diagnosis	Yes — with asymmetry L>R	Yes — with asymmetry R>L	Yes — without major asymmetry	Untestable
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)?	☐ 0	☐ 1	☐ 2	☐ 3	
A2.	Does the subject have limb weakness and/or hyperreflexia consistent with a diagnosis of primary lateral sclerosis (PLS) or ALS?	☐ 0	☐ 1	☐ 2	☐ 3	
A3.	Does the subject have bulbar weakness and/or fasciculations consistent with a diagnosis of ALS?	☐ 0			☐ 3	

Use the following criteria for 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.	

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[1] Brooks 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.

A4.	Does the subject have eye movement abnormalities consistent with a diagnosis of progressive supranuclear palsy (PSP)?	☐ 0		☐ 3	

Use the following criteria for A4, excerpted from SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders (Litvan et al., 2003)[1]:

NINDS-SPSP clinical criteria for the diagnosis of progressive supranuclear palsy (PSP)

Diagnostic categories	Inclusion criteria	Exclusion criteria	Supportive criteria
	For possible and probable: Gradually progressive disorder with age at onset at 40 or later;	For possible and probable: Recent history of encephalitis; alien limb syndrome; cortical sensory deficits; focal frontal or temporoparietal atrophy; hallucinations or delusions unrelated to dopaminergic therapy; cortical dementia of Alzheimer type; prominent, early cerebellar symptoms or unexplained dysautonomia; or evidence of other diseases that could explain the clinical features.	Symmetric akinesia or rigidity, proximal more than distal; abnormal neck posture, especially retrocollis; poor or absent response of parkinsonism to levodopa; early dysphagia & dysarthria; early onset of cognitive impairment including > 2 of: apathy, impairment in abstract thought, decreased verbal fluency, utilization or imitation behavior, or frontal release signs.
Possible	Either vertical supranuclear palsy or both slowing of vertical saccades and postural instability with falls < 1 yr disease onset.		
Probable	Vertical supranuclear palsy and prominent postural instability with falls within first year of disease onset.[a]		
Definite	All criteria for possible or probable PSP are met and histopathologic confirmation at autopsy.		

Adapted from Litvan I, Agid Y, Calne D et al. *Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report on the NINS-SPSP international workshop.* Nurology 1996:47:1-9.

[a] *Later defined as falls or the tendency to fall (patients are able to stabilize themselves).*

NINDS-SPSP = National Institute of Neurological Disorders and Stroke, and Society for Progressive Supranuclear Palsy, Inc

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[1] Litvan I, Bhatia KP, Burn DJ, et al. *Movement Disorders Society Scientific Issues Committee report: SIC Task force appraisal of clinical diagnostic criteria for Parkinsonian disorders.* Mov Disord. 2003 May; 18(5):467-86.

Questions A5–A9. Use the following criteria for A5–A9, excerpted from *Criteria for the diagnosis of corticobasal degeneration* (Armstrong et al., 2013)[1]. These criteria replace those of Lang and Kumar in use before 2013.

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

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

A5.	Does the subject have dystonia or rigidity consistent with a diagnosis of corticobasal degeneration (CBD)?	☐ 0	☐ 1 ☐ 2	☐ 3	

Dystonia and rigidity. Symptoms typically begin in one limb (no predilection for right or left side has been observed). Patients describe their limb difficulties as "clumsiness," "incoordination," or "stiffness." On examination, the limb is mildly to severely rigid and sometimes adopts a dystonic posture. Features of both rigidity (i.e., velocity-independent increased tone) and spasticity (i.e., velocity-dependent increased tone) can be present in the affected limbs. Dystonia in the head, neck, trunk or lower extremities is less common. The dystonia is often associated with rigidity, and the other signs, such as apraxia. The dystonic posture often involves the hand and forearm, with adduction of the shoulder, flexion of the fingers at the metacarophalangeal joints, and extension at the distal interphalangeal joints. Contractures are sometimes present. (From Thomas and Jankovic, *Parkinson Plus Syndromes* in Noseworthy, Neurological Therapeutics).

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

A6.	Is there ideomotor apraxia consistent with a diagnosis of CBD?	☐ 0	☐ 1 ☐ 2	☐ 3	☐ 8

Ideomotor apraxia: (including "limb kinetic aparaxia" and ideomotor apraxia) refers to the inability to translate an idea into a skilled motor act. The dysfunction cannot be explained by weakness or impaired comprehension. Motor apraxia (limb kinetic) and ideomotor apraxia are the types of limb apraxia relevant to this definition. Limb kinetic apraxia is tested with alternating motion tasks such as are performed in the UPDRS (UDS Form B3). Ideomotor apraxia of the arms is tested by asking the patient to mimic common intransitive gestures such as waving goodbye, hitchhiking, or saluting, or with transitive actions such as "show me how you would use a [comb, toothbrush, hammer, screwdriver]."

If ideomotor apraxia cannot be assessed at the neurological exam, please mark "Untestable" (8).

A7.	Is the alien limb phenomenon present consistent with a diagnosis of CBD?	☐ 0	☐ 1 ☐ 2		☐ 8

Alien limb phenomenon: Use the following definition[1]:

Patients often describe their affected limb as "alien," "uncontrollable," or "having a mind of its own," and they often label the limb "it" when describing the limb's behavior. The movements are spontaneous, minimally affected by mental effort, and sometimes requiring restraint by the contralateral limb. This phenomenon often lasts a few months to a few years before progressive rigidity or dystonia supercedes.

If alien limb phenomenon cannot be assessed at the neurological exam, please mark "Untestable" (8).

A8.	Is there myoclonus consistent with a diagnosis of CBD?	☐ 0	☐ 1 ☐ 2	☐ 3	

Myoclonus: a sudden shocklike twitching of muscles or parts of muscles without any rhythm or pattern.

Also use the following explanation for A8[1]:

Myoclonus, if present, usually begins distally in one upper limb and may spread proximally. The frequency and amplitude of myoclonic jerks typically increase with tactile stimulation (i.e., stimulus-sensitive myoclonus) and action (i.e., action myoclonus). Typically, a peripheral stimulus that induces myoclonic jerks is not associated with an enhanced somatosensory-evoked potential, and the latency from stimulus to jerk is brief, just sufficient to have reached the cortex and returned to the periphery (i.e., approximately 40 milliseconds in the upper limb). These features are distinct from most other forms of cortical reflex myoclonus (which is associated with enhanced somatosensory-evoked potential and a longer stimulus-to-jerk latency).

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[1] Boeve BF, Lang AE, Litvan I. C*orticobasal Degeneration and Its Relationship to Progressive Supranuclear Palsy and Frontotemporal Dementia.* Ann Neurol 2003; 54(Suppl 5):S15–S19.

A9.	Is there a cortical sensory deficit consistent with a diagnosis of CBD?	☐ 0	☐ 1 ☐ 2	☐ 3	☐ 8

Cortical sensory deficits are defined as impairments in the detection of complex sensory stimuli in the absence of elementary sensory deficits. This function is typically tested with number drawing in the palm or with asking the patient to identify coins placed between the thumb and index finger while the patient's eyes are closed. In the latter task, the patient is asked to manipulate the coin and then identify its denomination and which side is heads.

If cortical sensory deficit cannot be assessed at the neurological exam, please mark "Untestable" (8).

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.	Severity	☐ 0 Normal ☐ 1 Slight alteration in speed or fluidity of gait ☐ 2 Walks with difficulty but requires no assistance ☐ 3 Severe disturbance ☐ 4 Cannot walk at all ☐ 8 Untestable (specify reason): _____
B2.	Type	☐ 0 Normal ☐ 1 Hemiparetic (spastic) ☐ 2 Foot-drop gait (lower motor neuron) ☐ 3 Ataxic gait ☐ 4 Parkinsonian gait ☐ 5 Apractic gait ("magnetic gait") ☐ 6 Antalgic gait ☐ 7 Other gait disorder not listed above — specify: _____ ☐ 8 Untestable (specify reason): _____

The dominant gait disorder should be noted. If there are truly multiple gait disorders, use "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. This gait disorder is characterized in detail in the UPDRS (UDS Form B3). Note that if Parkinsonian gait is endorsed, the appropriate item in the standard UDS (UPDRS, Form B3, item 12) should also be abnormal.
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.** Use "7" for another 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, informant, medical records and/or observation. The neuropsychological test batteries and imaging results (both FTLD module and Uniform Data Set [UDS]) 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.

NOTE: This form is to be completed by a clinician with experience in evaluating subjects with frontotemporal lobar degeneration. For additional clarification, see FTLD Coding Guidebook for Follow-up Visit Packet, Form B9F. Check only one box per question.

Gateway question for primary progressive aphasia (PPA)

	No	Yes
1. **Does the subject have an acquired and progressive difficulty with language* consistent with PPA of a neurodegenerative type?** *DIFFICULTY WITH LANGUAGE: Other than simple dysarthria, are there difficulties with retrieving, using, repeating, sequencing, or understanding words? **If answer is "No", check "0 (Absent)" for Questions 2–11 and "0 (No)" for Question 12.**	☐ 0	☐ 1

If subject is a normal control, select "0 (No)."

TASKS THAT MAY BE USED TO ASSESS SPEECH AND LANGUAGE FUNCTIONS 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
Poor object naming. 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
Impoverished word selection/retrieval in spontaneous speech or writing. 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
Impaired word comprehension. 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
Poor object/person knowledge. This is different from not being able to name a person (anomia, item 2) or being unable to define a word (poor comprehension, item 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
Grammatical simplification or grammatical errors in speech or writing. 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, halting speech. 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.				
8. **Circumlocutory, empty speech** (Secondary diagnostic feature of logopenic variant; also present in semantic variant)	☐ 0	☐ 1	☐ 2	☐ 9

Characterizing speech and language symptoms / assigning PPA subtype

	Absent	Questionably present	Definitely present	Not evaluated
Circumlocutory speech, empty speech. 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)	☐ 0	☐ 1	☐ 2	☐ 9
Speech sound / word errors (paraphasias). 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)	☐ 0	☐ 1	☐ 2	☐ 9
Impaired speech repetition. 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 contect of the learning phase of a brief mental status examination's memory task.				
11. **Surface dyslexia and dysgraphia** — *also refer to Word Reading Test from FTLD Neuropsychological Battery* (Secondary feature of semantic variant)	☐ 0	☐ 1	☐ 2	☐ 9
Surface dyslexia and dysgraphia. 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?	☐ 0 No **Proceed to Question 14**		☐ 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 need to be met (see table on page 19). 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 1–2 years.				

Characterizing speech and language symptoms / assigning PPA subtype

	Absent	Questionably present	Definitely present	Not evaluated
13. **Consensus diagnosis of dominant PPA subtype based on clinician and neuropsychologist judgment** *NOTE: The diagnostic criteria in this module do not match the criteria in UDS V2.0 (Form D1). While Version 2.0 of the UDS is still in use, keep the two sets of diagnostic criteria separate.*	☐ 1 PPA, semantic variant (semPPA) ☐ 2 PPA, nonfluent/agrammatic variant (nf/gPPA) ☐ 3 PPA, logopenic variant ☐ 4 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.

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

Diagnostic features for the 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 2 of 3 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 3 must be present:

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

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

Diagnostic criteria for the 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 3 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 3 must be present:

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

Abbreviations: AD = Alzheimer's disease; FTLD = 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 3 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 3 must be present:

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

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

Gateway question for behavioral variant frontotemporal dementia (bvFTD)

	No	Yes
14. **Does the subject have acquired, clinically important alterations in behavior, personality, or comportment consistent with bvFTD of a neurodegenerative type?** **If answer is "No", check "0 (Absent)" for Questions 15–21 and "0 (No)" for Question 22.**	☐ 0	☐ 1

bvFTD gateway question. Question 14 asks the clinician whether there are prominent changes in behavior, personality or comportment 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.

"Of a neurodegenerative type" means that the etiology is not traumatic, vascular, etc.

If subject is a normal control, select "0 (No)."

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 comportment, and cognitive features that discriminate 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 3 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	☐ 0	☐ 1	☐ 2	☐ 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).

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
16. **Apathy or inertia** Loss of interest, drive, and motivation; decreased initiation of behavior	☐ 0	☐ 1	☐ 2	☐ 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.

17. **Loss of sympathy / empathy** Diminished response to other people's needs or feelings; diminished social interest, interrelatedness, or personal warmth	☐ 0	☐ 1	☐ 2	☐ 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	☐ 0	☐ 1	☐ 2	☐ 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	☐ 0	☐ 1	☐ 2	☐ 9

Hyperorality and dietary changes. 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 Kluver-Bucy syndrome.

20. **Changes on neuropsychological testing consistent with bvFTD** (refer to neuropsychological evaluation and neuropsychologist's impression)	☐ 0	☐ 1	☐ 2	☐ 9

Changes on neuropsychological testing. 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 comportment the principal cause of impaired daily living activities?	☐ 0	☐ 1	☐ 2	☐ 9

Impaired daily functioning. 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?** *PROBABLE: Meets three of the above criteria and has impaired daily functioning and has imaging consistent with bvFTD. **POSSIBLE: Meets three of the above criteria but is not functionally impaired or does not having imaging consistent with bvFTD *NOTE: The diagnostic criteria in this module do not match the criteria in UDS V2.0 (Form D1). While Version 2.0 of the UDS is still in use, keep the two sets of diagnostic criteria separate.*	☐ 0 Meets <3 of the features described in Questions 15–21: does not meet criteria for bvFTD; or an exclusionary feature is present. ☐ 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 diagnosis of bvFTD. This question is 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*

23 – 25. Self-explanatory questions regarding whether an electroyogram (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.**	☐ 0	☐ 1	
24. **Has an EMG been performed in the past year?** **If answer is "0 (No)", SKIP TO QUESTION 26.**	☐ 0	☐ 1	
25. **If an EMG was performed, did it show evidence of motor neuron disease?**	☐ 0	☐ 1	☐ 9

Check 9 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 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.

Check 9 only if the order of appearance is unknown.

Form C1F: Neuropsychological Battery Summary Scores

This form should be completed by Alzheimer's Disease Center (ADC) 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.

NOTE: 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 in the first data element and skip the rest of the data elements for that test:

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

1. Benson Complex Figure Copy

1a. Total score for copy of Benson figure (0–17) . . . . . ___ ___

The purpose of the test is to assess a subject's visuoconstructional and visual memory functions. In this test, the subject is presented with a figure composed of geometric shapes. The subject is then asked to reproduce the figure on the same page. The accuracy of each shape and its placement are recorded. The primary measure of performance is the total score for copying the Benson figure.

There may be instances when test administrators should consider the test invalid (e.g., if the subject did not bring his/her glasses and can't see well enough to take the test). In these instances, enter the appropriate code listed on Form C1F.

If a subject has motor problems and cannot complete the Benson Complex Figure Copy, a code of 95 (Physical problem) should be entered for the score on Form C1F, Q1a.

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.

2. Verbal Fluency: Phonemic Test

2a. Number of correct **F-words** generated in 1 minute (0–40) . . . . . ___ ___

2b. Number of **F-words** repeated in 1 minute (0–15) . . . . . ___ ___

2c. Number of **non-F-words** and rule violation errors in 1 minute (0–15) . . . . . ___ ___

2d. Number of correct **L-words** generated in 1 minute (0–40) . . . . . ___ ___

2e. Number of **L-words** repeated in one minute (0–15) . . . . . ___ ___

2f. Number of **non-L-words** and rule violation errors in 1 minute (0–15) . . . . . ___ ___

2g. TOTAL number of correct **F-words and L-words** (0–80) . . . . . ___ ___

2h. TOTAL number of **F-word and L-word** repetition errors (0–30) . . . . . ___ ___

2i. TOTAL number of **non-F/L words** and rule violation errors (0–30) . . . . . ___ ___

This is a widely used measure of word generation that may be sensitive to dysfunction in the dominant frontal lobe. In this test, the subject is told a letter of the alphabet (F) and asked to state as many words as possible that begin with that letter within 60 seconds. After 60 seconds, this is repeated with a second letter (L). The primary measure of performance is the total number of correct F-words and L-words.

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.

3. Word Reading Test — Regular and Irregular Words

REGULAR

3a. Total completely accurate words (0–15) . . . . . ___ ___

3b. Total semantically related inaccurate words (0–15) . . . . . ___ ___

3c. Total other phonologically related words or nonword errors (0–15) . . . . . ___ ___

IRREGULAR

3d. Total completely accurate words (0–15) . . . . . ___ ___

3e. Total semantically related inaccurate words (0–15) . . . . . ___ ___

3f. Total words that are "regularized" (read using "phonics," e.g., *sew* read as *sue*) (0–15) . . ___ ___

3g. 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.

4. Benson Complex Figure Delay (Recall)

4a. Total score for 10- to 15-minute delayed drawing of Benson figure (0–17) . . . . . ___ ___

4b. Recognized original stimulus from among four options? . . . . . ☐ 0 No ☐ 1 Yes

Approximately 10 to 15 minutes after the subject copies the Benson figure (see Benson Complex Figure Copy), the subject is asked to draw the figure again, by memory, on a blank page. The accuracy of each shape and its placement are recorded. The primary measure of performance is the total score for the 10- to 15-minute delayed drawing of the Benson figure.

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.

5. Semantic Word-picture Matching Test

5a. Total correct word-picture matches (0–20) . . . . . ___ ___

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.

6. Semantic Associates Test

6a. Total correct animal associations (0–8) . . . . . ___ ___

6b. Total correct tool associations (0–8) . . . . . ___ ___

6c. 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.

7. Northwestern Anagram Test — Short Form

7a. Correct subject who-questions (0–5) . . . . . ___ ___

7b. Correct object who-questions (0–5) . . . . . ___ ___

7c. 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.

8. Sentence Repetition Test

8a. Number of completely accurate sentences (0–5) . . . . . ___ ___

8b. Total number of words omitted from sentences (0–37) . . . . . ___ ___

8c. Total number of semantically related or unrelated incorrect real words (0–20) . . . . . ___ ___

8d. 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.

9. Noun and Verb Naming Subtests

9a. Total nouns correct (0–16) . . . . . ___ ___

9b. Total verbs correct (0–16). . . . . ___ ___

9c. Noun-to-verb ratio (total nouns correct / total verbs correct) . . . . . ___ ___ . ___ ___

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.

Reference: *Noun and Verb Naming Subtests from the Northwestern Naming Battery – Experimental Edition*. Copyright© 2011 by Cynthia K. Thompson and Sandra Weintraub. Reproduced by permission.

10. Sentence Reading Test

10a. Number of completely accurate sentences (0–5) . . . . . ___ ___

10b. Total number of words omitted from sentence (0–37) . . . . . ___ ___

10c. Total number of semantically related or unrelated incorrect real words (0–20) . . . . . ___ ___

10d. 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.

Form C2F: Social Norms Questionnaire[1]

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.

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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?	☐ 0 NO	☐ 1 YES
2.	Spit on the floor?	☐ 0 NO	☐ 1 YES
3.	Blow your nose in public?	☐ 0 NO	☐ 1 YES
4.	Ask a coworker their age?	☐ 0 NO	☐ 1 YES
5.	Cry during a movie at the theater?	☐ 0 NO	☐ 1 YES
6.	Cut in line if you are in a hurry?	☐ 0 NO	☐ 1 YES
7.	Laugh when you yourself trip and fall?	☐ 0 NO	☐ 1 YES
8.	Eat pasta with your fingers?	☐ 0 NO	☐ 1 YES
9.	Tell a coworker your age?	☐ 0 NO	☐ 1 YES
10.	Tell someone your opinion of a movie they haven't seen?	☐ 0 NO	☐ 1 YES
11.	Laugh when someone else trips and falls?	☐ 0 NO	☐ 1 YES
12.	Wear the same shirt every day?	☐ 0 NO	☐ 1 YES
13.	Keep money you find on the sidewalk?	☐ 0 NO	☐ 1 YES
14.	Pick your nose in public?	☐ 0 NO	☐ 1 YES
15.	Tell a coworker you think they are overweight?	☐ 0 NO	☐ 1 YES
16.	Eat ribs with your fingers?	☐ 0 NO	☐ 1 YES
17.	Tell a stranger you like their hairstyle?	☐ 0 NO	☐ 1 YES
18.	Wear the same shirt twice in two weeks?	☐ 0 NO	☐ 1 YES

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19.	Tell someone the ending of a movie they haven't seen?	☐ 0 NO	☐ 1 YES
20.	Hug a stranger without asking first?	☐ 0 NO	☐ 1 YES
21.	Talk out loud during a movie at the theater?	☐ 0 NO	☐ 1 YES
22.	Tell a coworker you think they have lost weight?	☐ 0 NO	☐ 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 Social Norms Questionnaire (SNQ22) SCORING KEY	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 Checklist[1]

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.

NOTE: This form is to be completed by the examiner who administered the neuropsychological 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:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Spontaneously mentioned that he/she was performing badly.	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Made other self-depreciatory comments	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Showed emotional distress over his/her performance / cognitive abilities	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
2.	**Showed too little self-consciousness / embarrassment for self:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Disrobed immodestly (took off shoes, belt, pants, etc.; lifted shirt, etc.)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Engaged in belching or flatulence, or picked nose without apology	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Giggled or otherwise made silly, childish comment or noise	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
3.	**Insensitive to others' embarrassment or privacy:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Insulted or made a negative comment about examiner	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Made an embarrassing comment about examiner	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Made an inappropriate or embarrassing joke	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+

4.	**Failed to adapt / defer to structure of testing situation established by examiner:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Resisted redirection while engaging in a verbal monologue	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Interrupted examiner	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Verbalized a desire to leave the evaluation prematurely	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	d. Physically attempted to leave the evaluation prematurely	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	e. Failed to maintain topic of discussion, initiated tangent	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 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.)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
5.	**Was preoccupied with time or kept a strict timetable:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Reminded examiner what time evaluation had to be finished	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	3 ☐ 4+
6.	**Acted overly dependent:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Mentioned caregiver's absence or asked when caregiver would return	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Asked for feedback on performance	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Showed frustration when examiner would not provide explicit feedback	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
7.	**Was anxious:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Mentioned being nervous about testing / performance anxiety	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Mentioned being nervous about diagnosis or prognosis	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
8.	**Was stimulus-bound:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Made stimulus-bound error on testing	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Picked up object on desk unnecessarily	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Circumstantial speech; overly focused on details, overly lengthy	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+

9.	**Was perseverative:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Repeated previous answer on testing	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Repeated an anecdote	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
10.	**Showed decreased initiation:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Began response in a notably delayed manner (not due to general slowing)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Required additional verbal prompting to initiate task	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 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):**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	3 ☐ **Severely**
	a. Lost task set / forgot instructions after performing task correctly	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Repeated rules to self multiple times during task	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Lost train of thought during conversation or response (demonstrated thought blocking)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
12.	**Was overly disclosing or inappropriately familiar:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
	a. Spontaneously revealed inappropriately personal information concerning self (only)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	b. Spontaneously revealed inappropriately personal information concerning a relative or friend (can also involve self)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	c. Stood or leaned too close to examiner (noticeably entered examiner's personal space)	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
	d. Touched examiner	☐ 0 Never	☐ 1 Once	☐ 2 2-3x	☐ 3 4+
13.	**Showed diminished social / emotional engagement:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 3 **Severely**
14.	**Showed exaggerated / labile emotional reactivity:**	☐ 0 **Not at all**	☐ 1 **A little bit**	☐ 2 **Moderately**	☐ 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.	___ ___ ___

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

[1]Reproduced by permission of the author, Katherine Rankin, PhD; do not copy or distribute without author's permission. Form created as part of the FTLD Module to the National Alzheimer's Coordinating Center Uniform Data Set, copyright 2013 University of Washington.

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 Scale[1] — INFORMANT QUESTIONNAIRE

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.

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, mark "Via telephone interview of informant by clinic staff". Check only one method.

THIS QUESTIONNAIRE WAS COMPLETED:

☐ 0 Independently by informant, as described in "Instructions to the Center"

☐ 1 Via in-person interview of informant by clinic staff

☐ 2 Via telephone interview of informant by clinic staff

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."	☐ 1	☐ 2	☐ 3	☐ 4
2. The subject worries about making mistakes.	☐ 1	☐ 2	☐ 3	☐ 4
3. Criticism or scolding hurts the subject quite a bit.	☐ 1	☐ 2	☐ 3	☐ 4
4. The subject feels pretty worried or upset when he/she thinks somebody is angry at him/her.	☐ 1	☐ 2	☐ 3	☐ 4
5. Even if something bad is about to happen to the subject, he/she rarely experiences fear or nervousness.	☐ 1	☐ 2	☐ 3	☐ 4
6. The subject feels worried when he/she thinks he/she has done poorly at something.	☐ 1	☐ 2	☐ 3	☐ 4
7. The subject has very few fears compared to his/her friends.	☐ 1	☐ 2	☐ 3	☐ 4

FOR CLINIC USE ONLY:

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:

  BIS1 + BIS2 + BIS3 + BIS4 + (5 – BIS5) + BIS6 + (5 – 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 Index[1] — 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.

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, mark "Via telephone interview of informant by clinic staff". Check only one method.

THIS QUESTIONNAIRE WAS COMPLETED:

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

PLEASE GIVE US SOME INFORMATION ABOUT YOURSELF:

Your sex: ☐ 1 Male ☐ 2 Female

Self explanatory.

Your date of birth (mm/yyyy): ___ ___ / ___ ___ ___ ___

The informant 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 informant 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").

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)

Self explanatory.

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
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.	☐1 ☐2 ☐3 ☐4 ☐5
9.	If the subject sees someone being treated unfairly, the subject doesn't feel much pity for him/her.	☐1 ☐2 ☐3 ☐4 ☐5
10.	The subject is often quite touched by things he/she sees happen.	☐1 ☐2 ☐3 ☐4 ☐5
11.	The subject believes that there are two sides to every question and tries to look at both of them.	☐1 ☐2 ☐3 ☐4 ☐5
12.	I would describe the subject as a pretty soft-hearted person.	☐1 ☐2 ☐3 ☐4 ☐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.	☐1 ☐2 ☐3 ☐4 ☐5
14.	Before criticizing me, the subject is likely to imagine how he/she would feel if he/she were in my place.	☐1 ☐2 ☐3 ☐4 ☐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): ___ ___

SCORING INSTRUCTIONS FOR FORM C5F

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

Form C6F: Revised Self-Monitoring Scale[1] — 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 Expressivness (EX) Score, the Ability to Modify Self-presentation (SP) Score, and the RSMS Total Score.

Indicate the method used to complete this questionnaire. If any of the questions were completed via telephone, mark "Via telephone interview of informant by clinic staff". Check only one method.

FOR CLINIC USE ONLY

THIS QUESTIONNAIRE WAS COMPLETED:

☐ 0 Independently by informant, as described in "Instructions to the Center"

☐ 1 Via in-person interview of informant by clinic staff

☐ 2 Via telephone interview of informant by clinic staff

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.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
2. The subject is often able to correctly read people's true emotions through their eyes.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 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.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 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.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
5. The subject's powers of intuition are quite good when it comes to understanding others.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
6. The subject can usually tell when others consider a joke in bad taste, even though they may laugh convincingly.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 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.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
8. The subject can usually tell when he/she said something inappropriate by reading it in the listener's eyes.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
9. The subject has trouble changing his/her behavior to suit different people and different situations.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
10. The subject can adjust his/her behavior to meet the requirements of any situation he/she is in.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
11. If someone is lying to the subject, he/she usually knows it at once from that person's manner or expression.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
12. Even when it might be to his/her advantage, the subject has difficulty putting up a good front.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5
13. Once the subject knows what the situation calls for, it's easy for him/her to regulate his/her actions accordingly.	☐ 0	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5

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.
- 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:

  RSMS2 + RSMS4 + RSMS5 + RSMS6 + RSMS8 + 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:

  RSMS1 + RSMS3 + RSMS7 + (5 – RSMS9) + RSMS10 + (5 – RSMS12) + 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:

  RSMS1 + RSMS2 + RSMS3 + RSMS4 + RSMS5 + RSMS6 + RSMS7 + RSMS8 + (5 – RSMS9) + RSMS10 + RSMS11 + (5 – RSMS12) + RSMS13

  If an item is missing, the total score should not be calculated. In this case, enter "88".
- Higher scores are interpreted as reflecting a greater degree of interpersonal sensitivity and responsiveness.

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).

NOTE: 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.	
1. If a subject has had a structural MRI scan, but the date and scan details have not yet been recorded on a FTLD module visit (IVP or FVP) as "available for data sharing," select Yes (1) and enter the information below.	
1a. Date of scan (MM / DD / YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	__ __ / __ __ / __ __ __ __
1a. Enter the month, day and year of the scan in the specified numerical format (e.g., March 1, 2010 would be entered as "03/01/2010"). If the exact day is unknown, enter '99' in the appropriate field (e.g., March 2010 would be 03/99/2010).	
1b. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
1b. Self explanatory.	
1c. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes ADNI version: _____ ☐ 9 Unknown
1c. 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
1d. Self explanatory.	
1d1. Scan model:	_____
1d1. If scan model is not known, enter "Unknown".	
1e. Field strength:	☐ 1 1.5T ☐ 2 3T ☐ 3 7T ☐ 4 Other: _____ ☐ 9 Unknown
1e. Self explanatory.	
1f. Are results of quantitative image analysis available?	☐ 0 No ☐ 1 Yes
1f. Self explanatory.	
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)	☐ 0 No ☐ 1 Yes
2. If a subject has had an FDG-PET scan, but the date and scan details have not yet been recorded on a FTLD module visit (IVP or FVP) as "available for data sharing," select Yes (1) and enter the information below.	
IF YES, complete 2a – 2e; if no, go to Question 3.	
2a. Date of scan (MM / DD / YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	__ __ / __ __ / __ __ __ __
2a. Enter the month, day, and year of the scan in the specified numerical format (e.g., March 1, 2010 would be entered as "03/01/2010"). If the exact day is unknown, enter '99' in the appropriate field (e.g., March 2010 would be 03/99/2010).	

2b. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
2b. Self explanatory.	
2c. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes *ADNI version:* _____ ☐ 9 Unknown
2c. For information on ADNI protocol, see Jagust WJ, Bandy D, Chen K, et al. *The Alzheimer's Disease Neuroimaging Initiative positron emission tomography core*. Alzheimers Dement 2010; 6(3):221-229.	
2d. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other: _____ ☐ 9 Unknown
2d. Self explanatory.	
2d1. Scan model:	_____
2d1. If scan model is not known, enter "Unknown".	
2e. Are results of quantitative image analysis available?	☐ 0 No ☐ 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)	☐ 0 No ☐ 1 Yes
3. If a subject has had an amyloid PET scan, but the date and scan details have not yet been recorded on a FTLD module visit (IVP or FVP) as "available for data sharing," select Yes (1) and enter the information below.	
IF YES, complete 3a – 3f; if no, go to Question 4.	

3a. Date of scan (MM / DD / YYYY): *NOTE: A value of 99 (unknown) is permissible for day only.*	__ __ / __ __ / __ __ __ __
3a. Enter the month, day, and year of the scan in the specified numerical format (e.g., March 1, 2010 would be entered as "03/01/2010"). If the exact day is unknown, enter '99' in the appropriate field (e.g., March 2010 would be 03/99/2010).	
3b. Is it in DICOM format or other electronic format?	☐ 0 No ☐ 1 Yes (specify format): _____ ☐ 9 Unknown
3b. Self explanatory.	
3c. Ligand used:	☐ 1 11C-PIB ☐ 2 18F-AV45 ☐ 3 Other (specify): _____ ☐ 9 Unknown
3c. Self explanatory.	
3d. Was ADNI protocol used?	☐ 0 No ☐ 1 Yes *ADNI version:* _____ ☐ 9 Unknown
3d. For information on ADNI protocol, see Jagust WJ, Bandy D, Chen K, et al. *The Alzheimer's Disease Neuroimaging Initiative positron emission tomography core.* Alzheimers Dement 2010; 6(3):221-229.	
3e. Scan manufacturer:	☐ 1 GE ☐ 2 Siemens ☐ 3 Philips ☐ 4 Other: _____ ☐ 9 Unknown
3e. Self explanatory.	
3e1. Scan model:	_____
3e1. If scan model is not known, enter "Unknown".	

3f. Are results of quantitative image analysis available?	☐ 0 No ☐ 1 Yes
3f. Self explanatory.	

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)	☐ 0 No ☐ 1 Yes *(If yes, identify type(s) below)*
4. If a subject has had other PET or SPECT scans, but the date and scan details have not yet been recorded on a FTLD module visit (IVP or FVP) as "available for data sharing," select Yes (1) and enter the information below.	
IF YES, complete 4a – 4d; if no, end form here.	
4a. Is a dopaminergic scan available?	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
4a. Self explanatory.	
4b. Is a serotonergic scan available?	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
4b. Self explanatory.	
4c. Is a cholinergic scan available?	☐ 0 No ☐ 1 Yes ☐ 9 Unknown
4c. Self explanatory.	
4d. Is another kind of scan available?	☐ 0 No ☐ 1 Yes (specify): _____
4d. Self explanatory.	

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).

NOTE: 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**	☐ 0	☐ 1	
1. Self explanatory.			
STRUCTURAL MRI			
2. Was structural MRI done? **If "No", SKIP TO QUESTION 3.**	☐ 0	☐ 1	
2. 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.**	☐ 0	☐ 1	☐ 9
2a. Check 9 only if focal atrophy status is unknown.			
Where was focal atrophy appreciated?			
2a1 – 2a11: Check 9 only if focal atrophy status is unknown in the specified location.			
2a1. Right frontal lobe	☐ 0	☐ 1	☐ 9
2a2. Left frontal lobe	☐ 0	☐ 1	☐ 9
2a3. Right temporal lobe	☐ 0	☐ 1	☐ 9
2a4. Left temporal lobe	☐ 0	☐ 1	☐ 9
2a5. Right medial temporal lobe	☐ 0	☐ 1	☐ 9
2a6. Left medial temporal lobe	☐ 0	☐ 1	☐ 9
2a7. Right parietal lobe	☐ 0	☐ 1	☐ 9

2a8. Left parietal lobe	☐ 0	☐ 1	☐ 9
2a9. Right basal ganglia	☐ 0	☐ 1	☐ 9
2a10. Left basal ganglia	☐ 0	☐ 1	☐ 9
2a11. Other area of the brain (specify below): _____	☐ 0	☐ 1	☐ 9
FDG-PET			
3. Was FDG-PET done? **If "No", SKIP TO QUESTION 4.**	☐ 0	☐ 1	
3. 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
3a. Check 9 only if focal atrophy status is unknown.			
Where was focal hypometabolism appreciated?			
3a1 – 3a11: Check 9 only if focal hypometabolism status is unknown in the specified location.			
3a1. Right frontal lobe	☐ 0	☐ 1	☐ 9
3a2. Left frontal lobe	☐ 0	☐ 1	☐ 9
3a3. Right temporal lobe	☐ 0	☐ 1	☐ 9
3a4. Left temporal lobe	☐ 0	☐ 1	☐ 9
3a5. Right medial temporal lobe	☐ 0	☐ 1	☐ 9
3a6. Left medial temporal lobe	☐ 0	☐ 1	☐ 9
3a7. Right parietal lobe	☐ 0	☐ 1	☐ 9
3a8. Left parietal lobe	☐ 0	☐ 1	☐ 9
3a9. Right basal ganglia	☐ 0	☐ 1	☐ 9
3a10. Left basal ganglia	☐ 0	☐ 1	☐ 9
3a11. Other area of the brain (specify below): _____	☐ 0	☐ 1	☐ 9

AMYLOID PET			
4. Was amyloid PET done? **If "No", SKIP TO QUESTION 5.**	☐ 0	☐ 1	
4. If amyloid PET was not done, skip this section.			
4a. Was amyloid deposition appreciated by visual inspection? **If "No" or "Unknown", SKIP TO QUESTION 5.**	☐ 0	☐ 1	☐ 9
4a. Check 9 only if amyloid deposition status is unknown.			
Where was amyloid deposition noted?			
4a1 – 4a11: Check 9 only if amyloid deposition status is unknown in the specified location.			
4a1. Right frontal lobe	☐ 0	☐ 1	☐ 9
4a2. Left frontal lobe	☐ 0	☐ 1	☐ 9
4a3. Right temporal lobe	☐ 0	☐ 1	☐ 9
4a4. Left temporal lobe	☐ 0	☐ 1	☐ 9
4a5. Right medial temporal lobe	☐ 0	☐ 1	☐ 9
4a6. Left medial temporal lobe	☐ 0	☐ 1	☐ 9
4a7. Right parietal lobe	☐ 0	☐ 1	☐ 9
4a8. Left parietal lobe	☐ 0	☐ 1	☐ 9
4a9. Right basal ganglia	☐ 0	☐ 1	☐ 9
4a10. Left basal ganglia	☐ 0	☐ 1	☐ 9
4a11. Other area of the brain (specify below): _____	☐ 0	☐ 1	☐ 9

	No	Yes	Unknown
CBF SPECT			
5. Was CBF SPECT done? **If "No", SKIP TO QUESTION 6.**	☐ 0	☐ 1	
5. If CBF SPECT was not done, skip this section.			
5a. Were abnormalities appreciated by visual inspection? **If "No" or "Unknown", SKIP TO QUESTION 6.**	☐ 0	☐ 1	☐ 9
5a. Check 9 only if the status of CBF SPECT-detected abnormalities is unknown.			
Where were abnormalities noted?			
5a1 – 5a11: Check 9 only if the status of CBF SPECT-detected abnormalities is unknown in the specified location.			
5a1. Right frontal lobe	☐ 0	☐ 1	☐ 9
5a2. Left frontal lobe	☐ 0	☐ 1	☐ 9
5a3. Right temporal lobe	☐ 0	☐ 1	☐ 9
5a4. Left temporal lobe	☐ 0	☐ 1	☐ 9
5a5. Right medial temporal lobe	☐ 0	☐ 1	☐ 9
5a6. Left medial temporal lobe	☐ 0	☐ 1	☐ 9
5a7. Right parietal lobe	☐ 0	☐ 1	☐ 9
5a8. Left parietal lobe	☐ 0	☐ 1	☐ 9
5a9. Right basal ganglia	☐ 0	☐ 1	☐ 9
5a10. Left basal ganglia	☐ 0	☐ 1	☐ 9
5a11. Other area of the brain (specify below): _____	☐ 0	☐ 1	☐ 9

	No	Yes	Unknown
OTHER IMAGING			
6. Was other imaging done?	☐ 0	☐ 1	
If yes, specify: _____			