FOLLOW-UP VISIT PACKET NACC UNIFORM DATA SET (UDS) — FTLD MODULE

Form A3F: Family History: Affected Family Members

Center: _____ Subject ID: _ _ _ _ _ _ _ _ _ _ Form Date: _ _/_ _/_ _ _ _

Visit #: _ _ _

Examiner's initials: _ _ _

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 Form A3F at all previous FTLD module visit(s), if applicable, with the subject or informant. For family members who were denoted as being affected at a previous FTLD module visit, any corrections to their data should be made to that 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.

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
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
1c. What is the predominant mutation?	☐ 1 MAPT ☐ 2 PGRN ☐ 3 C9ORF72 ☐ 4 FUS ☐ 8 Other (SPECIFY: _____) ☐ 9 Unknown		
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	___	___ ___ ___	___	___ ___ ___

***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 or UDS Follow-up 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 or UDS Follow-up Visit Form A3 with a birth year of 9999, then UDS Initial Visit or UDS Follow-up 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**

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****Codes for primary diagnosis**

See Appendix 1 on page 6 of this form

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

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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 child 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 or UDS Follow-up 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 or UDS Follow-up Visit Form A3 on FTLD Module Forms A3F and A3aF.**

If an affected child has already been listed on UDS Initial Visit or UDS Follow-up Visit Form A3 with a birth year of 9999, then UDS Initial Visit or UDS Follow-up 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.	__ __ / __ __ __ __	___	___ ___ ___	___	___ ___ ___

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7 Family report

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

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**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

***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 an 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.