

TELEPHONE FOLLOW-UP PACKET NACC UNIFORM DATA SET (UDS)

Form A3: Subject Family History

ADC name: _____ Subject ID: _____ Form date: ____ / ____ / _____
 Visit #: _____ Examiner's initials: _____

INSTRUCTIONS: This form is to be completed by a clinician with experience in evaluating patients with neurological problems and psychiatric conditions. For additional clarification and examples, see UDS Coding Guidebook for Telephone Follow-up Packet, Form A3.

SPECIAL INSTRUCTIONS for subjects who are receiving UDS Version 3 of Form A3 for the first time:

NOTE: A subject is receiving UDS v3 Form A3 for the first time if:

- No A3 data has been submitted yet for this subject –OR–
- A3 data has been submitted for this subject, but it was collected using UDS v2

For such subjects, you must fill out this form in its entirety, meaning:

1. You must answer **1=Yes** to Question 1 on genetic mutations and complete 2a – 4b.
2. You must answer **1=Yes** to Question 5 on parents and complete 5a – 5b.
3. You must answer **1=Yes** to Question 6a on siblings and complete 6aa – 6at, as appropriate.
4. You must answer **1=Yes** to Question 7a on children and complete 7aa – 7ao, as appropriate.

Corrections or new information on previously submitted family members — For family members who were denoted as being “affected” with a neurological or psychiatric condition or who were not affected at a previous UDS visit, any corrections to their data should be made to that previous A3 Form. Any newly obtained information (e.g., new mutation information, new diagnoses, new method of evaluation), including for family members previously reported as being affected at a past UDS visit, should be indicated on this form and should not be submitted as a correction to a previously submitted Form A3.

A summary of all previously submitted family history data can be found at: <https://www.alz.washington.edu/MEMBER/siteprint.html>.

1. Since the last visit, is new information available concerning genetic mutations addressed by Questions 2a through 4b, below?	<input type="checkbox"/> 0 No (SKIP TO QUESTION 5) <input type="checkbox"/> 1 Yes <input type="checkbox"/> 9 Unknown (SKIP TO QUESTION 5)
2a. In this family, is there evidence for an AD mutation? If Yes, select predominant mutation. NOTE: APOE should not be reported here.	<input type="checkbox"/> 0 No (SKIP TO QUESTION 3a) <input type="checkbox"/> 1 Yes, APP <input type="checkbox"/> 2 Yes, PS-1 (PSEN 1) <input type="checkbox"/> 3 Yes, PS-2 (PSEN 2) <input type="checkbox"/> 8 Yes, other (SPECIFY): _____ <input type="checkbox"/> 9 Unknown whether mutation exists (SKIP TO QUESTION 3a)
2b. Source of evidence for AD mutation (check one):	<input type="checkbox"/> 1 Family report (no test documentation available) <input type="checkbox"/> 2 Commercial test documentation <input type="checkbox"/> 3 Research lab test documentation <input type="checkbox"/> 8 Other (SPECIFY): _____ <input type="checkbox"/> 9 Unknown

<p>3a. In this family, is there evidence for an FTLD mutation? If Yes, select predominant mutation.</p>	<p><input type="checkbox"/> 0 No (SKIP TO QUESTION 4a)</p> <p><input type="checkbox"/> 1 Yes, MAPT</p> <p><input type="checkbox"/> 2 Yes, PGRN</p> <p><input type="checkbox"/> 3 Yes, C9orf72</p> <p><input type="checkbox"/> 4 Yes, FUS</p> <p><input type="checkbox"/> 8 Yes, other (SPECIFY): _____</p> <p><input type="checkbox"/> 9 Unknown whether mutation exists (SKIP TO QUESTION 4a)</p>
<p>3b. Source of evidence for FTLD mutation (check one):</p>	<p><input type="checkbox"/> 1 Family report (no test documentation available)</p> <p><input type="checkbox"/> 2 Commercial test documentation</p> <p><input type="checkbox"/> 3 Research lab test documentation</p> <p><input type="checkbox"/> 8 Other (SPECIFY): _____</p> <p><input type="checkbox"/> 9 Unknown</p>
<p>4a. In this family, is there evidence for a mutation other than an AD or FTLD mutation? (If No or Unknown, SKIP TO QUESTION 5)</p>	<p><input type="checkbox"/> 0 No (SKIP TO QUESTION 5)</p> <p><input type="checkbox"/> 1 Yes (SPECIFY): _____</p> <p><input type="checkbox"/> 9 Unknown (SKIP TO QUESTION 5)</p>
<p>4b. Source of evidence for other mutation (check one):</p>	<p><input type="checkbox"/> 1 Family report (no test documentation available)</p> <p><input type="checkbox"/> 2 Commercial test documentation</p> <p><input type="checkbox"/> 3 Research lab test documentation</p> <p><input type="checkbox"/> 8 Other (SPECIFY): _____</p> <p><input type="checkbox"/> 9 Unknown</p>

BIOLOGICAL PARENTS

5. Since the last UDS visit, is new information available concerning the status of the subject’s biological mother or father?

- 0 No (**SKIP TO QUESTION 6**) 1 Yes (**COMPLETE QUESTIONS 5A–5B, AS APPLICABLE**)

If birth year is unknown, please provide an approximate year on the Initial Visit Form A3 and ensure that it is consistently reported on all Forms A3 submitted (Initial Visit and Follow-up). If it is impossible for the subject and co-participant to estimate year of birth, enter 9999=Unknown. For any biological parent with a neurological or psychiatric problem, the entire row must be filled out. If the clinician cannot determine the primary neurological problem/psychiatric condition after reviewing all available evidence, enter 9=Unknown in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row. If the parent has no neurological or psychiatric problem, enter 8=N/A — *no neurological problem or psychiatric condition* in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row.

	Birth month/year (99/9999=Unknown)	Age at death (888=N/A, 999=Unknown)	Primary neurological problem/psychiatric condition*	Primary Dx**	Method of evaluation***	Age of onset (999=unknown)
			See CODES below this table			
5a. Mother	__ / ____	__	__	__	__	__
5b. 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, alcoholism, or depression
- 8 N/A — no neurological problem or psychiatric condition
- 9 Unknown

****CODES for primary diagnosis**

See Appendix 1 on page 5 of this form.

*****CODES for method of evaluation**

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

Year of birth for full siblings and biological children: If birth year is unknown, please provide an approximate year on UDS Initial Visit Form A3 and UDS Follow-up Visit Form A3 so that the sibling/child with unknown birth year ends up in correct birth order relative to the other siblings/children.

Example: A subject is the oldest of three children. The subject was born in 1940 and the middle sibling in 1943; the youngest sibling's birth year is unknown. An approximate birth year of 1944 or later should be assigned to the youngest sibling.

Use that same birth year on FTLD Module Form A3a, if applicable, and across all UDS visits so that any new information on a particular sibling or child can be linked to previously submitted information. If it is impossible for the subject and co-participant to estimate year of birth, enter 9999=Unknown.

FULL SIBLINGS

6. How many full siblings does the subject have? ____ If subject has no full siblings, **SKIP TO QUESTION 7.**

6a. Since the last UDS visit, is new information available concerning the status of the subject's siblings?

0 No (**SKIP TO QUESTION 7**) 1 Yes (**COMPLETE QUESTIONS 6aa–6at, AS APPLICABLE**)

For any full sibling with a neurological or psychiatric problem, the entire row must be filled out. If the clinician cannot determine the primary neurological problem/psychiatric condition after reviewing all available evidence, enter 9=Unknown in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row. If the sibling has no neurological or psychiatric problem, enter 8=N/A — *no neurological problem or psychiatric condition* in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row.

	Birth month/year (99/9999=Unknown)	Age at death (888 = N/A, 999 = unknown)	Primary neurological problem/psychiatric condition*	Primary Dx**	Method of evaluation***	Age of onset (999 = unknown)
See CODES on page 4						
6aa. Sibling 1	__/____	___	__	___	__	___
6ab. Sibling 2	__/____	___	__	___	__	___
6ac. Sibling 3	__/____	___	__	___	__	___
6ad. Sibling 4	__/____	___	__	___	__	___
6ae. Sibling 5	__/____	___	__	___	__	___
6af. Sibling 6	__/____	___	__	___	__	___
6ag. Sibling 7	__/____	___	__	___	__	___
6ah. Sibling 8	__/____	___	__	___	__	___
6ai. Sibling 9	__/____	___	__	___	__	___
6aj. Sibling 10	__/____	___	__	___	__	___
6ak. Sibling 11	__/____	___	__	___	__	___
6al. Sibling 12	__/____	___	__	___	__	___
6am. Sibling 13	__/____	___	__	___	__	___
6an. Sibling 14	__/____	___	__	___	__	___
6ao. Sibling 15	__/____	___	__	___	__	___
6ap. Sibling 16	__/____	___	__	___	__	___
6aq. Sibling 17	__/____	___	__	___	__	___
6ar. Sibling 18	__/____	___	__	___	__	___
6as. Sibling 19	__/____	___	__	___	__	___
6at. Sibling 20	__/____	___	__	___	__	___

BIOLOGICAL CHILDREN

7. How many biological children does the subject have? ____ If subject has no biological children, **END FORM HERE.**

7a. Since the last UDS visit, is new information available concerning the status of the subject's biological children?
 0 No (**END FORM HERE**) 1 Yes (**COMPLETE QUESTIONS 7aa–7ao, AS APPLICABLE**)

For any biological child with a neurological or psychiatric problem, the entire row must be filled out. If the clinician cannot determine the primary neurological problem/psychiatric condition after reviewing all available evidence, enter 9=Unknown in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row. If the child has no neurological or psychiatric problem, enter 8=N/A — *no neurological problem or psychiatric condition* in the **Primary neurological problem/psychiatric condition** column, and then skip the subsequent questions in the row.

	Birth month/year (99/9999=Unknown)	Age at death (888 = N/A, 999 = unknown)	Primary neurological problem/ psychiatric condition*	Primary Dx**	Method of evaluation***	Age of onset (999 = unknown)
See CODES below this table						
7aa. Child 1	__/____	____	__	____	__	____
7ab. Child 2	__/____	____	__	____	__	____
7ac. Child 3	__/____	____	__	____	__	____
7ad. Child 4	__/____	____	__	____	__	____
7ae. Child 5	__/____	____	__	____	__	____
7af. Child 6	__/____	____	__	____	__	____
7ag. Child 7	__/____	____	__	____	__	____
7ah. Child 8	__/____	____	__	____	__	____
7ai. Child 9	__/____	____	__	____	__	____
7aj. Child 10	__/____	____	__	____	__	____
7ak. Child 11	__/____	____	__	____	__	____
7al. Child 12	__/____	____	__	____	__	____
7am. Child 13	__/____	____	__	____	__	____
7an. Child 14	__/____	____	__	____	__	____
7ao. Child 15	__/____	____	__	____	__	____

***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, alcoholism, or depression
- 8 N/A — no neurological problem or psychiatric condition
- 9 Unknown

****CODES for primary diagnosis**

See Appendix 1 on page 5 of this form.

*****CODES for method of evaluation**

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

****APPENDIX 1: PRIMARY DIAGNOSIS CODES**

040	Mild cognitive impairment (MCI), not otherwise specified
041	MCI — single domain amnesic
042	MCI — multiple domain with amnesia
043	MCI — single domain nonamnesic
044	MCI — multiple domain nonamnesic
045	Impaired, but not MCI
050	Alzheimer's disease dementia
070	Dementia with Lewy bodies
080	Vascular cognitive impairment or dementia
100	Impairment due to alcohol abuse
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
140	Clinical progressive supranuclear palsy
150	Clinical corticobasal syndrome/corticobasal degeneration
160	Huntington's disease
170	Clinical 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's disease
240	Stroke
250	Hydrocephalus
260	Traumatic brain injury
270	CNS neoplasm
280	Other
310	Amyotrophic lateral sclerosis
320	Multiple sclerosis
999	Specific diagnosis unknown (<i>acceptable if method of evaluation is not by autopsy, examination, or dementia evaluation</i>)

Neuropathology diagnosis from autopsy

400	Alzheimer's disease neuropathology
410	Lewy body disease — neuropathology
420	Gross infarct(s) neuropathology
421	Hemorrhage(s) neuropathology
422	Other cerebrovascular disease neuropathology
430	ALS/MND
431	FTLD with Tau pathology — Pick's disease
432	FTLD with Tau pathology — CBD
433	FTLD with Tau pathology — PSP
434	FTLD with Tau pathology — argyrophillic grains
435	FTLD with Tau pathology — other
436	FTLD with TDP-43
439	FTLD other (FTLD-FUS, FTLD-UPS, FTLD NOS)
440	Hippocampal sclerosis
450	Prion disease neuropathology
490	Other neuropathologic diagnosis not listed above

*****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; and 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 co-participant 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 co-participant 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 cognitive impairment, brief formal cognitive testing should be included in the interview.

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's disease.

6. Subject and/or co-participant telephone interview

See definition No. 4 above.

7. Family report

Family report should be coded when the co-participant 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's disease.