**APPENDIX 1: PRIMARY DIAGNOSIS CODES

040	Mild cognitive impairment (MCI), not otherwise specified
041	MCI — single domain 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 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 *(acceptable if method of evaluation is not by autopsy, examination, or dementia evaluation)*

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 — argyrophyllic 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.