**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 — argyrophilic 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.