NACC Neuropathology (NP) Data Form

(Version 9.1, September 2008)

NOTE: Version 9 is NOT the most current version of the NP form and is not to be used for autopsies conducted on or after January 27, 2014. For the current version of the form, please visit http://www.alz.washington.edu.
# NACC Neuropathology Data Form

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<table>
<thead>
<tr>
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</thead>
<tbody>
<tr>
<td>1. MDS/UDS Patient ID</td>
<td></td>
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<tr>
<td>2. Date form completed</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Neuropath ID</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Gender</td>
<td></td>
<td>(M or F)</td>
</tr>
<tr>
<td>5. Age at death</td>
<td></td>
<td></td>
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<tr>
<td>6. Date of death</td>
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</tbody>
</table>

7. **Does the brain have any gross or microscopic pathology (including any Alzheimer type pathology such as senile plaques and neurofibrillary tangles)?**
   
   (mark one box)
   
   □ 1 Yes
   
   □ 2 No

(Note: For either response, items 8A through 24 must also be answered.)
Alzheimer’s Disease. For all brains in which there is any degree of Alzheimer type pathology (ranging from a few senile plaques and neurofibrillary tangles to advanced Alzheimer’s Disease), please indicate the nature of the pathology according to commonly used pathologic criteria.

8A. NIA/Reagan Institute neuropathological criteria used:

(mark one box)

- □ 1 High likelihood of dementia being due to Alzheimer’s disease
- □ 2 Intermediate likelihood of dementia being due to Alzheimer’s disease
- □ 3 Low likelihood of dementia being due to Alzheimer’s disease
- □ 4 Criteria not met
- □ 5 Not done
- □ 9 Missing/unknown

8B. CERAD neuropathological criteria used:

(mark one box)

- □ 1 Definite Alzheimer’s disease
- □ 2 Probable Alzheimer’s disease
- □ 3 Possible Alzheimer’s disease
- □ 4 Criteria not met
- □ 5 Not done
- □ 9 Missing/unknown

8C. ADRDA/Khachaturian neuropathological criteria used:

(mark one box)

- □ 1 Alzheimer’s disease
- □ 2 Criteria not met
- □ 3 Not done
- □ 9 Missing/unknown

8D. Other or unspecified neuropathological criteria used (e.g., Tierney, etc.):

(mark one box)

- □ 1 Alzheimer’s disease, unspecified
- □ 2 Criteria not met
- □ 3 Not done
- □ 9 Missing/unknown
Neurofibrillary Pathology. For all brains in which topographic staging of neurofibrillary degeneration was done, please indicate the stage with a number from 1–7. If Braak staging was not done, use number 8.

9. **Braak & Braak Neurofibrillary Stage.**
   
   *(mark one box)*
   
   - 1 Stage I
   - 2 Stage II
   - 3 Stage III
   - 4 Stage IV
   - 5 Stage V
   - 6 Stage VI
   - 7 Neurofibrillary degeneration not present
   - 8 Not assessed
   - 9 Missing/unknown

Plaque Score. For the most severely affected cortical region, please indicate the plaque score. Please use the Consortium to Establish a Registry of Alzheimer’s Disease (CERAD) standards for sparse, moderate, and frequent.

10. **Neuritic plaques (plaques with argyrophilic dystrophic neurites with or without dense amyloid cores).**
    
    *(mark one box)*
    
    - 1 Frequent neuritic plaques
    - 2 Moderate neuritic plaques
    - 3 Sparse neuritic plaques
    - 4 No neuritic plaques
    - 5 Not assessed
    - 9 Missing/unknown
### 11. Diffuse plaques (plaques with non-compact amyloid and no apparent dystrophic neurites).

* (mark one box)  
  - [ ] 1  Frequent diffuse plaques  
  - [ ] 2  Moderate diffuse plaques  
  - [ ] 3  Sparse diffuse plaques  
  - [ ] 4  No diffuse plaques  
  - [ ] 5  Not assessed  
  - [ ] 9  Missing/unknown

### 12. Is ischemic, hemorrhagic or vascular pathology present?

* (mark one box)  
  - [ ] 1  Yes  
  - [ ] 2  No  
  - [ ] 3  Not assessed  
  - [ ] 9  Missing/unknown  

(Note: Items 12A through 12L must also be answered.)

#### 12A. Are one or more large artery cerebral infarcts present?

* (mark one box)  
  - [ ] 1  Yes  
  - [ ] 2  No  
  - [ ] 3  Not assessed  
  - [ ] 9  Missing/unknown

#### 12B. Are one or more cortical microinfarcts (including “granular atrophy”) present?

* (mark one box)  
  - [ ] 1  Yes  
  - [ ] 2  No  
  - [ ] 3  Not assessed  
  - [ ] 9  Missing/unknown

CONTINUE with 12C on the next page.
12C. Are one or more lacunes (small artery infarcts and/or hemorrhages) present?

(mark one box)

- □ 1 Yes
- □ 2 No
- □ 3 Not assessed
- □ 9 Missing/unknown

12D. Are single or multiple hemorrhages present?

(mark one box)

- □ 1 Yes
- □ 2 No
- □ 3 Not assessed
- □ 9 Missing/unknown

12E. Is subcortical arteriosclerotic leukoencephalopathy present?

(mark one box)

- □ 1 Yes
- □ 2 No
- □ 3 Not assessed
- □ 9 Missing/unknown

12F. Is cortical laminar necrosis present?

(mark one box)

- □ 1 Yes
- □ 2 No
- □ 3 Not assessed
- □ 9 Missing/unknown

12G. Is medial temporal lobe sclerosis (including hippocampal sclerosis) present?

(mark one box)

- □ 1 Yes
- □ 2 No
- □ 3 Not assessed
- □ 9 Missing/unknown

*CONTINUE with 12H on the next page.*
12H. Is atherosclerotic vascular pathology (of the circle of Willis) present?
   (mark one box)
   □ 1  None
   □ 2  Mild
   □ 3  Moderate
   □ 4  Severe
   □ 5  Not assessed
   □ 9  Missing/unknown

12I. Is arteriosclerosis (small parenchymal arteriolar disease) present?
   (mark one box)
   □ 1  None
   □ 2  Mild
   □ 3  Moderate
   □ 4  Severe
   □ 5  Not assessed
   □ 9  Missing/unknown

12J. Is amyloid angiopathy present?
   (mark one box)
   □ 1  None
   □ 2  Mild
   □ 3  Moderate
   □ 4  Severe
   □ 5  Not assessed
   □ 9  Missing/unknown

12K. Is another type of angiopathy (e.g., CADASIL or arteritis) present?
   (mark one box)
   □ 1  Yes
   □ 2  No
   □ 3  Not assessed
   □ 9  Missing/unknown

CONTINUE with 12L on the next page.
12L. Is there other pathology related to ischemic or vascular disease not previously specified present?

(mark one box)

☐ 1 Yes
☐ 2 No
☐ 3 Not assessed
☐ 9 Missing/unknown

Lewy Body Pathology. For all brains in which Lewy bodies are detected, indicate the presence and distribution of Lewy-related pathology. This classification is independent of the clinical presentation, which may be variable and include Parkinsonism, dementia, psychosis, sleep disorders, etc.

13A. Pathology is consistent with criteria of Consortium on Dementia with Lewy Bodies for:

(select only one)

☐ 1 Lewy body pathology, brainstem predominant type
☐ 2 Lewy body pathology, intermediate or transitional (limbic) type
☐ 3 Lewy body pathology, diffuse (neocortical) type
☐ 4 Lewy body pathology, unspecified or not further assessed
☐ 5 No Lewy bodies
☐ 6 Not assessed
☐ 9 Missing/unknown

13B. Likelihood that DLB Clinical Syndrome due to DLB Pathology:

(select only one)

☐ 1 Low
☐ 2 Intermediate
☐ 3 High
☐ 6 N/A (not applicable)
☐ 9 Missing/unknown
**Frontotemporal Degenerations (FTD).** Use this for non-Alzheimer degenerative disorders that commonly have the brunt of cortical changes in frontal and temporal lobes, but may involve other cortical and subcortical regions and have variable clinical presentations, including frontal lobe dementia, progressive aphasia, progressive apraxia, etc.

14A. Pick's Disease:

(mark one box)

- 1  Yes
- 2  No
- 3  Not assessed
- 9  Missing/unknown

14B. Corticobasal degeneration:

(mark one box)

- 1  Yes
- 2  No
- 3  Not assessed
- 9  Missing/unknown

14C. Progressive supranuclear palsy:

(mark one box)

- 1  Yes
- 2  No
- 3  Not assessed
- 9  Missing/unknown

14D. Frontotemporal dementia and Parkinsonism with tau-positive or argyrophilic inclusions:

(mark one box)

- 1  Yes
- 2  No
- 3  Not assessed
- 9  Missing/unknown

CONTINUE with 14E on the next page.
14E. Tauopathy, other (e.g., tangle-only dementia and argyrophilic grain dementia):
   (mark one box)
   □ 1 Yes
   □ 2 No
   □ 3 Not assessed
   □ 9 Missing/unknown

14F. FTD with ubiquitin-positive (tau-negative) inclusions:
   (mark one box)
   □ 1 FTD with motor neuron disease
   □ 2 FTD without motor neuron disease
   □ 3 None present
   □ 4 Not assessed
   □ 9 Missing/unknown

14G. Is there FTD with no distinctive histopathology (tau-negative, ubiquitin-negative, and no argyrophilic inclusions)?
   (mark one box)
   □ 1 Yes
   □ 2 No
   □ 3 Not assessed
   □ 9 Missing/unknown

14H. Was FTD “not otherwise specified” present (e.g., “immunostaining for ubiquitin and tau not done”)?
   (mark one box)
   □ 1 Yes
   □ 2 No
   □ 3 Not assessed
   □ 9 Missing/unknown
Prion-related Disorders:
15A. Is Creutzfeldt-Jakob disease or variant CJD present?
   (mark one box)
   □ 1  Yes
   □ 2  No
   □ 3  Not assessed
   □ 9  Missing/unknown

15B. Are other prion diseases present (e.g., Gerstmann-Straussler syndrome)?
   (mark one box)
   □ 1  Yes
   □ 2  No
   □ 3  Not assessed
   □ 9  Missing/unknown

Other Major Pathologic Disorders (e.g., infectious, immunologic, metabolic, neoplastic, toxic or degenerative).
16A. Are other major pathologic disorders present (not addressed by questions 8–15)?
   (mark one box)
   □ 1  Yes
   □ 2  No
   □ 3  Not assessed
   □ 9  Missing/unknown

   SKIP: If 2, 3 or 9, go to #17A.

16B. If 16A is yes, specify below (one disorder per line):

1 __________________________________________________________________________

2 __________________________________________________________________________

3 __________________________________________________________________________
17A. **Family history information relevant to neuropathologic diagnosis.** Choose one of the following categories that most accurately describes the family information available:

(mark one box)

- □ 1 Family history of similar neurodegenerative disorder
- □ 2 Family history of other (dissimilar) neurodegenerative disorder
- □ 3 No family history of similar or dissimilar neurodegenerative disorder
- □ 4 Family history of both similar and dissimilar neurodegenerative disorder
- □ 9 Family history unknown/not available/missing

**SKIP:** If 1, 3 or 9, go to #18A.

17B. If 17A is 2 or 4, specify disorder: _____________________________________________

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**Genetic Variants or Polymorphisms.** For each of the following three common genetic variants or polymorphisms, choose the patient’s genotype, if known; select “not available or not assessed” if unknown:

18A. **Apolipoprotein-E:**

(mark one box)

- □ 1 e3, e3
- □ 2 e3, e4
- □ 3 e3, e2
- □ 4 e4, e4
- □ 5 e4, e2
- □ 6 e2, e2
- □ 9 Missing/unknown/not assessed

18B. **Tau haplotype:**

(mark one box)

- □ 1 H1, H1
- □ 2 H1, H2
- □ 3 H2, H2
- □ 4 Other polymorphism (e.g., A0)
- □ 9 Missing/unknown/not assessed

*CONTINUE with 18C on the next page.*
18C. PRNP codon 129:
(mark one box)
- 1 M, M
- 2 M, V
- 3 V, V
- 9 Missing/unknown/not assessed

19. Genetic or chromosomal abnormalities. Choose below the one known genetic or chromosomal abnormality that best describes the subject:
(mark one box)
- 1 APP mutation
- 2 PS1 mutation
- 3 PS2 mutation
- 4 Tau mutation
- 5 α-Synuclein mutation
- 6 Parkin mutation
- 7 PRNP mutation
- 8 Huntingtin mutation
- 9 Notch 3 mutation (CADASIL)
- 10 Other known genetic mutation (e.g., ABri, neuroserpin)
- 11 Down syndrome
- 12 Other chromosomal abnormality
- 13 No known genetic or chromosomal abnormality
- 50 Not assessed
- 99 Missing/unknown
20. What are the primary and contributing pathologic diagnoses or features which you judge to be responsible for the subject’s cognitive status?

NOTE: Mark only one diagnosis as “primary”; any number may be marked as “contributing”.

<table>
<thead>
<tr>
<th>Primary</th>
<th>Contributing</th>
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<tbody>
<tr>
<td>A1</td>
<td>A2</td>
</tr>
<tr>
<td>B1</td>
<td>B2</td>
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<td>C1</td>
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<td>J2</td>
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<tr>
<td>K1</td>
<td>K2</td>
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**Brain Tissue and Post Mortem CSF.** Use this section to record information related to the storage and accessibility of brain tissue and post mortem CSF at your Center.

21. Is banked frozen brain tissue accessible?

   (mark one box)

   - [ ] 1 Yes
   - [ ] 2 No

22. Is formalin-fixed brain tissue accessible?

   (mark one box)

   - [ ] 1 Yes
   - [ ] 2 No
<table>
<thead>
<tr>
<th>Question</th>
<th>Options</th>
</tr>
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<tbody>
<tr>
<td>Are paraffin-embedded blocks of brain tissue accessible?</td>
<td>Yes, No</td>
</tr>
<tr>
<td>Is banked postmortem cerebrospinal fluid (CSF) accessible?</td>
<td>Yes, No</td>
</tr>
</tbody>
</table>