

**Supplemental Table 1. Symptoms in the DIAN cohort, with UDS terms and descriptions**

| Symptom                               | Section  | Description  |
|---------------------------------------|--|--|
| <b>Parkinsonism</b>                   | UDS B9: Clinician Judgment of Symptoms   | 12a. If there were changes in motor function, were these suggestive of parkinsonism?   |
| <b>Myoclonus</b>                      | UDS B3: UPDRS; Neurological Exam   | Mentioned specifically in commentary   |
| <b>Seizures</b>                       | UDS A5: Subject Health History   | a. Seizures (recent/active only)   |
| <b>Spasticity</b>                     | UDS B3: UPDRS; Neurological exam   | Mentioned specifically in commentary   |
| <b>Corticobulbar deficits</b>         | UDS B3: UPDRS; Neurological Exam   | Mentioned specifically in commentary   |
| <b>Cerebellar ataxia</b>              | UDS B3: UPDRS<br>Neurological Exam   | Mentioned specifically in commentary<br>Impaired finger-nose-finger or heel-shin, excluding instances of limited comprehension, limb weakness, or confounding apraxia  |
| <b>Aphasia</b>                        | UDS B9: Clinician Judgment of Symptoms   | 4c. Language (For example, does s/he have hesitant speech; have trouble finding words; use inappropriate words without self-correction?)   |
| <b>Apraxia</b>                        | UDS B3: UPDRS; Neurological exam   | Mentioned specifically in commentary   |
| <b>Visual agnosia</b>                 | UDS B9: Clinician Judgment of Symptoms   | 4d. Visuospatial function (Difficulty interpreting visual stimuli and finding his/her way around.)   |
| <b>Hallucinations</b>                 | UDS B9: Clinician Judgment of Symptoms<br>UDS B9: Clinician Judgment of Symptoms   | 7c. 1) Psychosis: Visual hallucinations<br>7c. 2) Psychosis: Auditory hallucinations   |
| <b>Behavioral/personality changes</b> | UDS B9: Clinician Judgment of Symptoms<br>UDS B9: Clinician Judgment of Symptoms<br>UDS B9: Clinician Judgment of Symptoms<br>UDS B9: Clinician Judgment of Symptoms | 7d. Disinhibition (Does the subject use inappropriate coarse language or exhibit inappropriate speech or behaviors in public or in the home? Does s/he talk personally to strangers or have disregard for personal hygiene?)<br>7e. Irritability (Does the subject overreact, such as shouting at family members or others?)<br>7f. Agitation (Does the subject have trouble sitting still; does s/he shout, hit, and/or kick?)<br>7g. Personality change (Does the subject exhibit bizarre behavior or behavior uncharacteristic of the subject, such as unusual collecting, suspiciousness [without delusions], unusual dress, or dietary changes? Does the subject fail to take other's feelings into account?) |
| <b>Stroke</b>                         | UDS B2: HIS and CVD  | 6. History of stroke (recent/active only)  |