**Supplementary Material**

**Family History Is Associated with Phenotype in Dementia with Lewy Bodies**

**Supplementary Figure 1.** Distribution of different groups according to family history. Family members with PDD were assigned to the subcategory ‘Family history of dementia & PD’, and family members with DLB were assigned to the subcategory ‘Family history of dementia’. DLB, dementia with Lewy bodies; PD, Parkinson’s disease; PDD, Parkinson’s disease dementia.

**Supplementary Table 1.** Differences in demographic features, clinical features and biomarkers between DLB patients with either a family history of dementia or a family history of PD.

|  |  |  |  |
| --- | --- | --- | --- |
|  | Family history of dementia only  (*n* = 69) | Family history of PD only  (*n* = 15) | *p* |
| Study center |  |  | 0.72 |
| Elisabeth-TweeSteden Hospital | 11 (16%) | 3 (20%) |  |
| Erasmus Medical Center | 7 (10%) | 2 (13%) |  |
| Amsterdam University Medical Center | 51 (74%) | 10 (67%) |  |
| Sex, male | 55 (80%) | 13 (87%) | 0.73 |
| Age of onset, y | 64.6 (8.0) | 64.5 (8.7) | 0.99 |
| First symptom  Cognitive decline  Parkinsonism  Hallucinations | 60 (87%)  4 (6%)  5 (7%) | 10 (67%)  5 (33%)  0 (0%) | 0.12  0.008\*  0.58 |
| MTA score (average of right and left) (n=51;9) | 1 (0.5-1.5) | 1 (0-1.3) | 0.24a |
| CSF tau/Aβ42 ratio >0.52 (n=43;6) | 22 (51%) | 1(17%) | 0.13a |
| *APOE* ε4 carrier (n=43;6) | 28 (65%) | 4 (67%) | 1.00 |

Values are presented as mean (SD), median (IQR) or *n* (%). DLB, dementia with Lewy bodies; PD, Parkinson’s disease; CSF, cerebrospinal fluid; MTA, medial temporal lobe atrophy, Aβ42, amyloid-β1-42, acorrected for time between age of onset and date of MRI or date of lumbar puncture, \**p*<0.05.

**Supplementary Table 2.** Statistical models regarding rate of cognitive decline and survival in the group of DLB patients with either a family history of dementia or a family history of PD.

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **Rate of cognitive decline** |  | β | SE |  | 95% CI | *p* |  |
| Uncorrected | Baseline | 0.67 | 1.51 |  | -2.33-3.66 | 0.66 |  |
| (Linear mixed model analysis) | Change over time | -0.022 | 0.045 |  | -0.11-0.067 | 0.62 |  |
|  |  |  |  |  |  |  |  |
| With correction for sex, age of onset, and study center  (Linear mixed model analysis) | Baseline | 0.34 | 1.50 |  | -2.63-3.31 | 0.82 |  |
|  | Change over time | -0.015 | 0.045 |  | -0.10-0.074 | 0.73 |  |
| **Survival** |  | Mediana | SE |  | 95% CI | *p* |  |
| Uncorrected  (Kaplan-Meier analysis) | Family history of dementia only | 10.0 | 0.65 |  | 8.72-11.28 | 0.97 |  |
|  | Family history of PD only | 10.0 | 1.98 |  | 6.12-13.88 |  |
|  |  |  |  |  |  |  |  |
|  | | B | SE | HR | 95% CI | *p* | Z-score |
| With correction for sex, age of onset, and study center  (Cox regression analysis) |  | 0.20 | 0.46 | 1.23 | 0.50-3.03 | 0.66 | 2.17 |

a Time between age of onset and death, \**p*<0.05.