

**eTable 1. Sensitivity analysis for comparisons between baseline characteristics of included (n=113) and not included (no follow-up data available) participants (n=26).**

|                                          | <b>not included</b> | <b>included</b> | <b>p-value</b> |
|------------------------------------------|---------------------|-----------------|----------------|
| <b>No. (%) of patients</b>               | 26 (100)            | 113 (100)       |                |
| <b>Definite/Probable CAA-ri, No. (%)</b> | 16 (61.5)           | 93 (82.3)       | 0.03           |
| <b>Possible CAA-ri, No. (%)</b>          | 10 (38.5)           | 20 (17.7)       |                |
| <b>Female, No. (%)</b>                   | 17 (65.4)           | 49 (43.4)       | 0.04           |
| <b>Age at onset, mean (SD), y</b>        | 71.0 (8.3)          | 72.9 (7.2)      | 0.28           |
| <b>&lt; 70 y, No. (%)</b>                | 8 (30.8)            | 32 (28.3)       | 0.80           |
| <b>History of</b>                        |                     |                 |                |
| MCI or AD dementia                       | 3 (11.5)            | 41 (36.3)       | 0.01           |
| ICH                                      | 5 (19.2)            | 38 (33.6)       | 0.15           |
| <b>APOEε4, No. (%)</b>                   |                     |                 |                |
| Non carriers                             | 4/13 (30.8)         | 61/97 (62.9)    | 0.01           |
| Heterozygote                             | 8/13 (61.5)         | 22/97 (22.7)    |                |
| Homozygote                               | 1/13 (7.7)          | 14/97 (14.4)    |                |
| <b>Symptoms onset, No. (%)</b>           |                     |                 |                |
| Cognitive/Behavioral Changes             | 14 (53.6)           | 81 (71.7)       | 0.08           |
| Focal Deficits                           | 7 (26.9)            | 65 (57.5)       | 0.01           |
| Seizures                                 | 3 (11.5)            | 39 (34.5)       | 0.02           |
| Headache                                 | 3 (11.5)            | 25 (22.1)       | 0.23           |
| Number of symptoms ≤1                    | 17 (65.4)           | 42 (37.2)       | 0.01           |
| <b>CMBs count, No. (%)</b>               |                     |                 |                |
| None <sup>a</sup>                        | -                   | 3 (2.7)         | 0.51           |
| 1                                        | 1 (3.9)             | 8 (7.1)         |                |
| 2 - 4                                    | 5 (19.2)            | 10 (8.9)        |                |
| 5 - 10                                   | 6 (23.1)            | 24 (21.2)       |                |
| >10                                      | 14 (53.9)           | 68 (60.1)       |                |
| <b>cSS</b>                               | 7 (27.0)            | 57 (50.4)       | 0.03           |
| <b>cSS+CMBs</b>                          | 7 (26.9)            | 54 (47.8)       | 0.05           |
| <b>Gd contrast-enhanced MRI, No. (%)</b> | 9/11 (81.8)         | 59/93 (63.4)    | 0.23           |

**eTable 1 Legend.**

Abbreviations: AD: Alzheimer's disease; MCI: Mild Cognitive Impairment; ICH: Intracerebral hemorrhage; APOEε4: Apolipoprotein ε4 allele; CMBs: cerebral microbleeds; cSS: cortical superficial siderosis; Gd: gadolinium; MRI: Magnetic Resonance Imaging. Unless otherwise indicated, data are expressed as number or mean (percentage or SD) referred to the total number of patients of each column. <sup>a</sup> all the three patients had cSS.