**Cerebrovascular disease progression in patients with *ACTA2* Arg179 Pathogenic Variants**

**Supplementary materials:**

**Supplementary tables:**

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| S1. Longitudinal Evaluation of Imaging Markers on Identical Scanner Setups (n=11) |
|  | Baseline | Last Follow-Up | p value |
| WMH, median cc [IQR] | 28.01 [13.57 – 58.71] | 25.95 [14.98 – 57.62] | 0.998 |
| CL, median no. [IQR] | 3 [0.25 – 13.25] | 9 [3.75 – 20.0] | 0.014 |
| CS, median no. [IQR] | 0.5 [0.0 – 2.0] | 2.5 [0.75 – 4.0] | 0.031 |
| Rel. ICA-Stenosis, mean (SD) | 0.36 (0.16) | 0.29 (0.09) | 0.033 |

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| S2: Characteristics of female vs. male ACTA2 Arginine 179 patients |
|  | Female (n=20) | Male (n=7) | p value |
| Age at baseline, y, median [range] | 2.8 [0.005-26.21] | 1.4 [0.014-14.1] | 0.42 |
| Age at last presentation, y, median [range] | 6.1 [0.030-32.47] | 7.2 [0.014-18/73] | 0.76 |
| p.Arg179His, n (%) | 14 (70.0) | 5 (83.3) | 1.00 |
| p.Arg179Cys, n (%) | 6 (30.0) | 1 (16.7) | 0.63 |
| WMH, median cc [IQR] | 25.95 [15.33-52.21] | 40.23 [10.80-86.21] | 0.89 |
| AIS, median no. [IQR] | 0 [0-16] | 0 [0-6] | 0.13 |
| CL, median no. [IQR] | 6.0 [0-28] | 0.0 [0-11.5] | 0.14 |
| CS, median no. [IQR] | 0 [0-4] | 0 [0-2] | 0.69 |
| mCASS, median no. [IQR] | 8 [3-21] | 3 [1-8] | 0.19 |
| Rel. ICA-Stenosis, mean (SD) | 0.34 (0.16) | 0.43 (0.17) | 0.26 |

**Supplementary table legends:**

**Supplementary Table 1 Abbreviations:**

WMH= white matter hyperintensities; AIS= ischemic stroke related to large artery occlusion; CL = cystic-like white matter lesions; CS = critical stenosis on TOF MRA, mCASS = modified Focal **c**erebral **a**rteriopathy of **c**hildhood **S**everity **S**core; Rel. ICA-Stenosis = terminal to the petrous ICA diameter.

**Supplementary Table 2 Abbreviations:** WMH= white matter hyperintensities; AIS= ischemic stroke related to large artery occlusion; CL = cystic-like white matter lesions; CS = critical stenosis on TOF MRA, mCASS = modified Focal **c**erebral **a**rteriopathy of **c**hildhood **S**everity **S**core; Rel. ICA-Stenosis = terminal to the petrous ICA diameter. Denominators may vary slightly due to missing data regarding mutation in one patient. Data are reported as median, raw quantiles, mean with standard deviations or raw frequencies with weighted percentages.