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

Supplementary materials:

Supplementary tables:

e-1: 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

e-2: 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:

Table e-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.

Table e-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.