

Supplementary Table 1. Clinical and neuroimaging features of CADASIL patients in different H-Y stages

	H-Y stage 0 (n=20)	H-Y stage 1–2 (n=40)	H-Y stage 3–5 (n=15)	P
Age (yr)	57.0 (53.0–61.5)	65.0 (58.0–69.5)	74.0 (71.0–80.0)	<0.01
Male sex	8 (40.0)	20 (50.0)	11 (73.3)	0.14
UPDRS part III	0 (0–0)	9 (6.5–14.5)	31 (26–40)	<0.01
H-Y stage	0 (0–0)	1 (1–2)	3 (3–4)	<0.01
Hypertension	10 (50.0)	25 (62.5)	11 (73.3)	0.39
Diabetes mellitus	2 (10.0)	14 (35.0)	2 (13.3)	0.07
Dyslipidemia	10 (50.0)	21 (52.5)	6 (40.0)	0.75
Smoking	0 (0.0)	13 (32.5)	5 (33.3)	<0.01
Stroke	9 (45.0)	25 (62.5)	11 (73.3)	0.25
α -synuclein (pg/mL)	0.12 (0.07–0.18)	0.08 (0.07–0.13)	0.09 (0.07–0.13)	0.64
Gait speed (m/s)	1.06 (0.96–1.26)	0.78 (0.66–1.06)	0.33 (0.13–0.57)	<0.01
Cortical thickness (mm)	2.39 (2.36–2.46)	2.37 (2.29–2.44)	2.19 (2.13–2.31)	<0.01
WMH volume (mL)	20.2 (7.9–40.1)	42.0 (25.1–57.7)	56.0 (42.1–65.7)	<0.01
Fazekas scale				
Periventricular	2 (1–3)	3 (2–3)	3 (2–3)	0.02
Subcortical	3 (3–3)	3 (2–3)	3 (3–3)	<0.01
Total	6 (5–6)	6 (5–6)	6 (5–6)	<0.01
Basal ganglia dilated PVS	3 (2–3)	3 (2–3)	3 (2–4)	0.20
Lacune number	3 (2–9)	5 (2.5–11)	10 (5–15)	0.05
Any BG lacune	6 (30.0)	23 (57.5)	10 (66.7)	0.07
BG lacune number	0 (0–1)	1 (0–2)	1 (0–3)	0.11
CMB number	11 (1–23)	12 (3–58)	39 (9–76)	0.06
Any BG CMB	8 (40.0)	23 (57.5)	12 (80.0)	0.07
BG CMB number	0 (0–4)	0 (1–4.5)	2 (1–5)	0.33

Values are presented as median (interquartile range) or n (%) unless otherwise indicated.

CADASIL, cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy; UPDRS, Unified Parkinson's Disease Rating Scale; H-Y, Hoehn-Yahr; WMH, white matter hyperintensity; PVS, perivascular spaces; CMB, cerebral microbleeds; BG, basal ganglia.