

Supplementary Table 3. Genotype–phenotype correlation of the *RNF213* R4810K variant in pediatric moyamoya disease

Variable	GG (n=9)	GA (n=49)	AA (n=5)	P	Post-hoc P		
					GG vs. GA	GG vs. AA	GA vs. AA
Female sex	2 (22)	32 (65)	1 (20)	0.01	0.08	>0.99	0.07
Age at onset (yr)	9.0 [7.8, 9.0]	7.7 [6.2, 11.6]	4.3 [1.9, 9.5]	0.60			
Age under 5	1 (11)	7 (14)	3 (60)	0.04	>0.99	0.28	0.11
Family history	0 (0)	8 (16)	0 (0)	0.51			
Revascularization surgery	8 (89)	45 (92)	3 (60)	0.14			
Clinical manifestation							
Infarction	0 (0)	5 (10)	2 (40)	0.14			
TIA	4 (44)	34 (69)	1 (20)	0.05			
ICH/IVH	0 (0)	2 (4)	0 (0)	>0.99			
Seizure	2 (22)	3 (6)	2 (40)	0.03	0.50	>0.99	0.06
Incidental	0 (0)	0 (0)	0 (0)	-			
Others*	3 (33)	5 (10)	0 (0)	0.15			
Angiographic findings							
Bilateral vasculopathy	7 (78)	42 (86)	5 (100)	0.67			
PCA involvement	2 (22)	7 (14)	4 (80)	<0.001	>0.99	0.27	<0.001
Suzuki grade [†]				0.72			
1–2	1 (25)	3 (18)	0 (0)				
3–4	2 (50)	12 (71)	2 (67)				
5–6	1 (25)	2 (12)	1 (33)				

Data are presented as n (%) or median [interquartile range].

RNF213, ring finger protein 213; GG, wild type; GA, heterozygote; AA, homozygote; TIA, transient ischemic attack; ICH/IVH, intracranial cerebral hemorrhage/ intraventricular hemorrhage; PCA, posterior cerebral artery.

*Others: headache, dizziness, and syncope; [†]The Suzuki grade on the severe side was applied.