

Supplementary Table 4. Comparison between pediatric and adult moyamoya disease among *RNF213* R4810K heterozygotes

Variable	Pediatric (n=49)	Adult (n=189)	P
Female	32 (65)	121 (64)	0.87
Family history	8 (16)	36 (19)	0.66
Revascularization surgery	45 (92)	35 (19)	<0.001
Clinical manifestation			
TIA	34 (69)	52 (28)	<0.001
Infarction	5 (10)	39 (21)	0.09
ICH/IVH	2 (4)	21 (11)	0.14
Seizure	3 (6)	3 (2)	0.07
Incidental	0 (0)	40 (21)	<0.001
Others*	5 (10)	34 (18)	0.19
Angiographic findings			
Bilateral vasculopathy	42 (86)	113 (60)	0.001
PCA involvement	7 (14)	41 (22)	0.25
Suzuki grade [†]			0.65
1–2	3 (18)	40 (26)	
3–4	12 (71)	96 (62)	
5–6	2 (12)	20 (13)	

Data are presented n (%).

RNF213, ring finger protein 213; 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.

Supplementary Table 6. Characteristic of patients with revascularization surgery

	Number of hemispheres	Number of patients
Revascularization surgery	99	65
Direct	11	10
Indirect	88	55
<i>RNF213</i> genotype		
Wild type (GG)	11	8
Heterozygote (GA)	85	55
Homozygote (AA)	3	2

RNF213, ring finger protein 213.

Supplementary Table 5. Clinical Outcomes in patients with moyamoya disease and the R4810K variant of *RNF213*

Variable	All (n=293)	GG (n=64)	GA (n=225)	AA (n=4)	P
Infarction	30 (10.2)	4 (6.3)	24 (10.7)	2 (50.0)	0.04
TIA	51 (17.4)	11 (17.2)	39 (17.3)	1 (25.0)	0.80
ICH/IVH	11 (3.8)	2 (3.1)	8 (36.7)	1 (25.0)	0.16
Death	2 (0.7)	1 (1.6)	1 (0.4)	0 (0.0)	0.41
FU period (mo)	34.9 [21.2–61.7]	29.3 [20.9–47.4]	36.8 [21.0–64.7]	53.5 [26.9–148.2]	0.13

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; FU, follow-up.