

**Supplemental material**

**Title**

Isolated anterior cerebral artery occlusion: an atypical form of moyamoya disease

**Journal name**

*Stroke and Vascular Neurology*

**Table S1.** Comparison of the primary symptoms and genotype in adult and pediatric patients

<b>First admission</b>	<b>Age &lt; 18 years old (n=27)</b>	<b>Age ≥ 18 years old (n=112)</b>	<b>P value</b>
<b>Female</b>	14(51.9%)	65(58.0%)	0.560
<b>Primary symptom at onset</b>			
<b>Asymptomatic</b>	0	1(0.9%)	1.000
<b>Transient ischemic attack</b>	19 (70.4%)	68(60.7%)	0.352
<b>Infarction</b>	3(11.1%)	12(10.7%)	1.000
<b>Haemorrhage</b>	4(14.8%)	20(18.3%)	0.881
<b>Headache</b>	1(3.7%)	5(4.6%)	1.000
<b>Others</b>	0	6(5.5%)	0.599
<b>Mutation †</b>			0.066
<b>GG</b>	16/25(64.0%)	74/91(81.3%)	
<b>GA</b>	9/25(36.0%)	17/91(18.7%)	

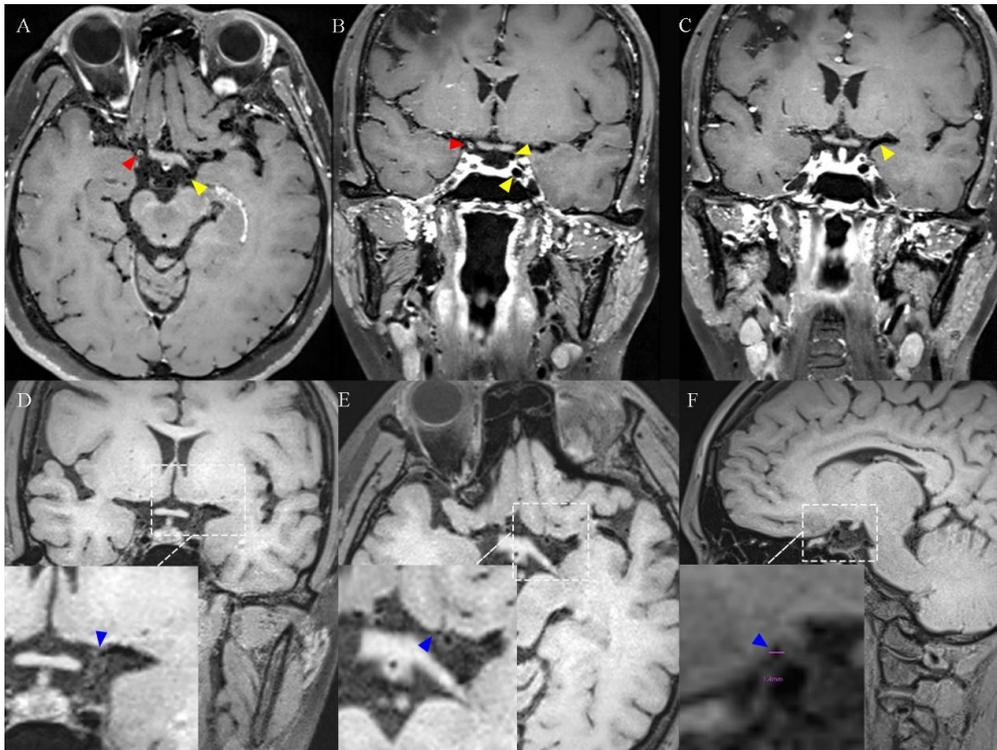
GG, wildtype *RNF213* p.R4810K; GA, heterozygous *RNF213* p.R4810K; ACA, anterior cerebral artery; PCA, posterior cerebral artery; mRS, modified Rankin scale; MMD, moyamoya disease.

†*RNF 213* p.R4810K (rs112735431) gene mutation analysis was performed in 116 of the 139 patients

**Table S2.** Neuropsychological test performances of patients with ACA-occlusive MMD

	Number (n=65)	%
<b>MoCA (Mean±SD)</b>	21.82±4.21	
<10	0	0
10–17	10	15.4
18–26	37	56.9
≥26	18	27.7
<b>IADL (Mean±SD)</b>	14.48±1.89	
<16	59	90.8
16–22	5	7.7
≥22	1	1.5
<b>HAMD-17 (Mean±SD)</b>	4.37±3.80	
<7	49	75.4
7–17	16	24.6
18–24	0	0
>24	0	0

MoCA, Montreal Cognitive Assessment; IADL, Instrumental Activity of Daily Living Scale; HAMD, 17-item Hamilton Rating Scale for Depression.



**Figure S1.** The radiological characteristics of high-resolution MRI in ACA-occlusive MMD patients. a-c shows the HR-MRI diagram of the ICA in ACA-occlusive MMD. The red arrows indicate a centripetal constriction of the distal segment of the internal carotid artery in the contralateral hemisphere, while the yellow arrows indicate that there is usually no significant lesion in the ipsilateral ICA and MCA. d-f shows the HR-MRI image of ACA occlusion. The blue arrows indicates a centripetal constriction at the proximal section of the ACA. The outer diameter of the ACA here is 1.40 mm. MRI, magnetic resonance imaging; HR, high resolution; ACA, anterior cerebral artery; MMD, moyamoya disease; ICA, internal carotid artery.