

Table1. Demographic, clinical and radiographic characteristics, and cognitive profiles

Patient characteristics	Whole group (N=30)	Stroke presentation		p-value
		Stroke group (N=17)	No stroke group (N=13)	
Age at presentation (Mean±SD, Median, Range)	8.37 ± 3.44, 8.21, 15.13	7.66 ± 3.35, 7.90, 14.37	9.28 ± 3.47, 9.08, 11.99	0.21
Age at diagnosis, years (Mean±SD, Median, Range)	9.13 ± 3.18, 8.69, 12.29	8.72 ± 3.24, 8.12, 11.45	9.66 ± 3.13, 9.72, 11.04	0.43
Age at scan, years (Mean±SD, Median, Range)	12.55 ± 3.03, 12.62, 10.65	12.92 ± 3.45, 14.20, 10.65	12.06 ± 2.43, 12.18, 7.94	0.45
Females (N, %)	17 (56.7)	11 (64.7)	6 (46.2)	0.31
Ethnicity (N, %)				-
Caucasian	12 (40.0)	7 (53.8)	5 (29.4)	
African	3 (10.0)	0 (0.0)	3 (17.6)	
East-Asian	10 (33.3)	4 (30.8)	6 (35.3)	
South-East Asian	1 (3.3)	0 (0.0)	1 (5.9)	
South-African	4 (13.3)	2 (15.4)	2 (11.8)	
Moyamoya classification (%)				
Idiopathic	15 (50.0)	9 (52.9)	6 (46.2)	0.71
Syndromic	15 (50.0)	8 (47.1)	7 (53.8)	
NF1	12 (80.0)	5 (29.5)	7 (53.8)	-
SCD	3 (20.0)	3 (17.6)	0 (0.0)	-
Clinical presentation (%)				
Stroke	17 (56.7)	17 (100.0)	0 (0.0)	-
TIA	9 (30.0)	4 (23.5)	5 (38.5)	0.38
Headache	6 (20.0)	4 (23.5)	2 (15.4)	0.58
Asymptomatic	10 (33.3)	6 (35.3)	4 (30.8)	0.79
Other	2 (6.7)	0 (0.0)	2 (15.4)	0.25
Radiographic findings (%)				
Parenchymal				
Cortical Watershed	17 (56.7)	17 (56.7)	0 (0.0)	-
MCA-ACA	16 (53.3)	16 (53.3)	0 (0.0)	-
PCA-ACA	3 (10.0)	3 (10.0)	0 (0.0)	-
Deep watershed	6 (20.0)*	4 (13.3)*	2 (6.6)**	-
Suzuki grade (%)				
Left hemisphere				
0	7 (23.3)	3 (17.6)	4 (30.8)	0.23
1	0 (0.0)	0 (0.0)	0 (0.0)	
2	6 (20.0)	6 (35.3)	0 (0.0)	
3	15 (50.0)	7 (41.2)	8 (61.5)	
4	2 (6.7)	1 (5.9)	1 (7.7)	
Right hemisphere				
0	6 (20.0)	1 (5.9)	5 (38.5)	0.04

	1	1 (3.3)	1 (5.9)	0 (0.0)	
	2	1 (3.3)	0 (0.0)	1 (7.7)	
	3	17 (56.7)	12 (70.6)	5 (38.5)	
	4	5 (16.7)	3 (17.6)	2 (15.4)	
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Vascular (%)					
Moyamoya laterality					
	Bilateral	17 (56.7)	13 (76.5)	4 (30.8)	0.06
	Left	6 (20.0)	1 (5.9)	5 (38.5)	
	Right	7 (23.3)	3 (17.6)	4 (30.8)	
Intellectual ability (Wechsler Intelligence Quotient) (Mean ± SD)					
	FSIQ (Full-scale IQ)	89.87 ± 15.31	87.06 ± 17.70	93.54 ± 11.10	0.26
	VCI (Verbal Comprehension Index)	91.53 ± 13.50	86.88 ± 13.77	97.62 ± 10.81	0.03
	PRI (Perceptual Reasoning Index)	91.23 ± 18.21	88.65 ± 19.74	95.23 ± 15.58	0.36
	WMI (Working Memory Index)	93.07 ± 13.57	93.82 ± 16.64	92.08 ± 8.60	0.71
	PSI (Processing Speed Index)	92.13 ± 16.50	92.06 ± 20.03	92.23 ± 11.07	0.98
Executive function (parent-reported BRIEF) (Mean ± SD)					
	Behavior Regulation/ Inhibitory Self-Control Index (BRIS)	53.27 ± 13.01 (36 - 81)	55.08 ± 13.76	53.50 ± 12.79	0.77
	Metacognitive Index (MI)	58.83 ± 14.03 (34 - 84)	53.82 ± 10.49	62.36 ± 15.17	0.14
	Global Executive composite (GEC)	57.83 ± 13.17 (35 - 77)	55.18 ± 12.02	60.45 ± 14.33	0.36

* Cortical infarct (1/4), ** one lacunar infarct and one caudate, SD: standard deviation, NF1: Neurofibromatosis type 1, SCD: Sickle cell disease, TIA: Transient ischemic attack, ACA: Anterior cerebral artery, MCA: Middle cerebral artery, PCA: Posterior cerebral artery

Table 2. Descriptive statistics of fractional voxel counts of negatively reacting voxels

		HM	No stroke group			Stroke group		
			Mean±SD	Median	Range	Mean±SD	Median	Range
Brain tissue	Grey matter	L	0.19 ± 0.12	0.14	0.05 - 0.39	0.19 ± 0.11	0.17	0.06 - 0.41
		R	0.15 ± 0.10	0.11	0.05 - 0.39	0.20 ± 0.09	0.17	0.08 - 0.32
	White matter	L	0.21 ± 0.11	0.21	0.07 - 0.38	0.23 ± 0.12	0.21	0.10 - 0.49
		R	0.18 ± 0.10	0.13	0.05 - 0.36	0.22 ± 0.10	0.20	0.09 - 0.38
Brain region	Frontal	L	0.24 ± 0.20	0.19	0.06 - 0.69	0.27 ± 0.12	0.27	0.08 - 0.54
		R	0.22 ± 0.21	0.16	0.05 - 0.78	0.25 ± 0.13	0.22	0.06 - 0.47
	Parietal	L	0.16 ± 0.12	0.17	0.04 - 0.43	0.20 ± 0.15	0.14	0.06 - 0.57
		R	0.12 ± 0.06	0.12	0.04 - 0.22	0.18 ± 0.11	0.16	0.04 - 0.43
	Temporal	L	0.17 ± 0.09	0.16	0.05 - 0.38	0.20 ± 0.12	0.17	0.06 - 0.45
		R	0.12 ± 0.04	0.12	0.06 - 0.19	0.17 ± 0.09	0.15	0.06 - 0.40
	Occipital	L	0.12 ± 0.13	0.07	0.02 - 0.49	0.09 ± 0.06	0.08	0.03 - 0.26
		R	0.09 ± 0.08	0.07	0.02 - 0.24	0.11 ± 0.09	0.08	0.02 - 0.33

Table 3. Descriptive statistics for cognitive outcomes

Cognitive measures	Mean \pm SD, Median, Range	One sample t-test <i>p</i> -value	% Children with impaired function
Wechsler Intelligence Quotient (mean=100, SD=15)			≤ 85 (1SD)
FSIQ (Full-scale IQ)	89.87 \pm 15.31, 92.50, 48 – 111	0.001	36.7%
VCI (Verbal Comprehension Index)	91..53 \pm 13.50, 95.00, 56 – 115	0.002	26.7%
PRI (Perceptual Reasoning Index)	91.23 \pm 18.21, 94.25, 52 – 119	0.017	40.0%
WMI (Working Memory Index)	93.07 \pm 13.57, 94.00, 62 – 117	0.009	26.7%
PSI (Processing Speed Index)	92.13 \pm 16.50, 92.50, 53 – 126	0.014	33.3%
BRIEF Executive Function (parent form) (Mean=50, SD=10)			≥ 60 (1SD)
Behavior Regulation/ Inhibitory Self-Control Index (BRIS)	53.27 \pm 13.01, 51.50, 36 - 81	0.120	29%
Metacognitive Index (MI)	58.83 \pm 14.03, 60.50, 34 – 84	0.005	41%
Global Executive composite (GEC)	57.83 \pm 13.17, 58.50, 35 – 77	0.008	35.5%

Table 4. Relative effects of regional negative CVRs and cognitive outcomes

Cognitive outcomes		Effect	B	SE	Beta	95% CI		$p_{\text{corrected}}$	R^2
						LL	UL		
IQ	FSIQ	Intercept	128.63	8.65		110.82	146.45	<0.001	0.58
		Comorbidity	-17.73	4.80	-0.59	-27.62	-7.85	0.001	
		Left MM	-16.18	5.19	-0.46	-26.86	-5.49	0.005	
		Right MM	-13.85	5.62	-0.37	-25.43	-2.28	0.021	
		Right occipital lobe	-64.07	25.51	-0.36	-116.60	-11.54	0.038	
	VCI	Intercept	106.33	3.95		98.21	114.45	<0.001	0.42
		Stroke	-10.25	4.03	-0.38	-18.53	-1.96	0.017	
		Comorbidity	-5.85	4.33	-0.22	-14.75	3.05	0.188	
		Right occipital lobe	-60.58	25.79	-0.38	-113.60	-7.56	0.027	
	PRI	Intercept	99.21	4.44		90.09	108.33	<0.001	0.20
Comorbidity		-15.96	6.28	-0.45	-28.86	-3.07	0.017		
WMI	Intercept	109.71	4.78		99.91	119.51	<0.001	0.36	
	Comorbidity	-8.73	4.13	-0.33	-17.20	-0.27	0.044		
	Left white matter	-56.51	18.66	-0.47	-94.80	-18.22	0.005		
PSI	Intercept	108.49	5.86		96.46	120.51	<0.001	0.28	
	Comorbidity	-12.34	5.31	-0.38	-23.23	-1.45	0.028		
	Left frontal lobe	-39.79	17.31	-0.38	-75.30	-4.28	0.029		
EF	BRIS	Intercept	43.57	4.13		35.01	52.13	<0.001	0.31
		Right parietal lobe	70.81	22.78	0.55	23.57	118.06	0.005	
	MI	Intercept	47.36	5.61		35.67	59.06	<0.001	0.19
		Right white matter	55.06	25.37	0.44	2.14	107.98	0.056	
	GEC	Intercept	46.56	5.40		35.30	57.82	<0.001	0.22
	Right white matter	57.79	24.44	0.47	6.81	108.77	0.028		