

SUPPLEMENTAL MATERIAL

Supplemental Table I. Demographics and clinical characteristics of 355 childhood arterial ischemic stroke cases, stratified by primary arteriopathy diagnosis

Characteristic	Definite Arteriopathy N=127		Possible Arteriopathy N=34		No Arteriopathy N=194		P-value*
	n	(%)	n	(%)	n	(%)	
Demographics							
Age in years, median (IQR)	7.4	(3.7, 11.9)	9.1	(4.3, 15.7)	8.3	(2.1, 14.8)	0.31**
Male gender	68	53.5	22	64.7	109	56.2	0.51
Race							0.39
White	77	(60.6)	20	(58.8)	133	(68.6)	
Black	15	(11.8)	6	(17.6)	18	(9.3)	
Indian/South Asian	15	(11.8)	2	(5.9)	9	(4.6)	
East Asian	4	(3.1)	1	(2.9)	4	(2.1)	
First Nations/Aboriginal	2	(1.6)	0	(0)	2	(1.0)	
Middle Eastern	0	(0)	0	(0)	3	(1.5)	
Pacific Islander	0	(0)	0	(0)	1	(0.5)	
Mixed or other	11	(8.7)	4	(11.8)	23	(11.9)	
Unknown	3	(2.4)	1	(2.9)	1	(0.5)	
Ethnicity							0.07
Non-Hispanic	110	(86.6)	30	(88.2)	149	(76.8)	
Hispanic	10	(7.9)	2	(5.9)	35	(18.0)	
Mixed or other	7	(5.5)	2	(5.9)	10	(5.2)	
Country							0.05
USA	77	(60.6)	22	(64.7)	124	(63.9)	
Canada	23	(18.1)	5	(14.7)	32	(16.5)	
Australia	8	(6.3)	2	(5.9)	6	(3.1)	
Philippines	11	(8.7)	1	(2.9)	4	(2.1)	
Chile	2	(1.6)	0	(0)	12	(6.2)	
United Kingdom	2	(1.6)	1	(2.9)	8	(4.1)	
France	4	(3.1)	1	(2.9)	1	(0.5)	
Serbia	0	(0)	1	(2.9)	4	(2.1)	
China	0	(0)	1	(2.9)	3	(1.5)	
Stroke presentation							
Focal signs							
Hemiparesis	104	(81.9)	25	(73.5)	156	(80.4)	0.55
Dysarthria	33	(26.0)	12	(35.3)	52	(26.8)	0.54
Aphasia	35	(27.6)	5	(14.7)	39	(20.1)	0.16
Ataxia	27	(21.3)	13	(38.2)	31	(16.0)	0.004
Visual field deficit	15	(11.8)	7	(20.6)	24	(12.4)	0.38
Non-focal signs							
Headache	48	(37.8)	15	(44.1)	63	(32.5)	0.50
Decreased level of consciousness	37	(29.1)	12	(35.3)	53	(27.3)	0.83

Nausea/vomiting	31 (24.4)	9 (26.5)	45 (23.2)	0.96
Seizures at presentation	32 (25.2)	6 (17.6)	46 (23.7)	0.02
Vertigo	13 (10.2)	5 (14.7)	22 (11.3)	0.93
Diplopia	2 (1.6)	2 (5.9)	8 (4.1)	0.66
Papilledema	3 (2.4)	0 (0)	1 (0.5)	0.55
Risk factors or co-morbidities (not mutually exclusive)				
Cardiac disease	14 (11.0)	8 (23.5)	85 (43.8)	<0.0001
Congenital heart disease	6 (4.7)	2 (5.9)	56 (28.9)	<0.0001*
Acquired heart disease	0 (0)	1 (2.9)	20 (10.3)	<0.0001*
Isolated patent foramen ovale	6 (4.7)	4 (11.8)	11 (5.7)	0.28*
Stroke at cardiac surgery (<72 hours)	1 (0.8)	0 (0)	9 (4.6)	0.10*
Other cardiac disease	5 (3.9)	4 (11.8)	32 (16.5)	0.001*
Other chronic disorders				
Sickle cell anemia	9 (7.1)	0 (0)	4 (2.1)	0.05*
Downs syndrome	7 (5.5)	2 (5.9)	2 (1.0)	0.03*
Other genetic syndrome	5 (3.9)	1 (2.9)	10 (5.2)	0.93
Migraine	5 (3.9)	0 (0)	7 (3.6)	0.75*
Prothrombotic state	2 (1.6)	0 (0)	8 (4.1)	0.38*
Oral contraceptives***	3 (5.1)	2 (16.7)	5 (5.9)	0.28*
Indwelling catheter	1 (0.8)	0 (0)	8 (4.1)	0.15*
Iron deficiency anemia	4 (3.1)	1 (2.9)	1 (0.5)	0.09*
Brain tumor	4 (3.1)	0 (0)	1 (0.5)	0.17*
Benign	2 (1.6)	0 (0)	0 (0)	0.31*
Malignant	2 (1.6)	0 (0)	1 (0.5)	0.68*
Aneurysm	1 (0.8)	0 (0)	2 (1.0)	1.00*
PHACES syndrome/hemangioma	2 (1.6)	0 (0)	1 (0.5)	0.68*
Hematologic malignancy	0 (0)	0 (0)	2 (1.0)	0.61
L-asparaginase therapy	0	0 (0)	2 (1.0)	0.61*
Connective tissue disease	1 (0.8)	1 (2.9)	0 (0)	0.08*
Ventriculoperitoneal shunt	1 (0.8)	0 (0)	0 (0)	0.45*
Acute systemic illness by chart review				
Fever lasting >48 hours	18 (14.2)	2 (5.9)	24 (12.4)	0.47*
Systemic sepsis or bacteremia	9 (7.1)	2 (5.9)	9 (4.6)	0.65*
Dehydration	6 (4.7)	1 (2.9)	11 (5.7)	0.94*
Shock	0 (0)	1 (2.9)	8 (4.1)	0.04*
Viral gastroenteritis	0 (0)	0 (0)	2 (1.0)	0.61*

*P-value calculated using Fishers exact test (indicated by *); otherwise, chi-square tests were used

** Kruskal-Wallis test

***females only: N=59 (definite), 12 (possible), 85 (no arteriopathy)

Supplemental Table II. Imaging modality and timing of vascular imaging in 355 childhood arterial ischemic stroke cases, stratified by primary arteriopathy diagnosis

	Total N=355 n (%)	Definite Arteriopathy n %	Possible Arteriopathy n %	No Arteriopathy n %	P-value
Imaging modality at baseline					
Brain parenchyma					0.47
MRI only	216 (60.8)	72 (56.7)	20 (58.8)	124 (63.9)	
CT only	5 (1.4)	1 (0.8)	1 (2.9)	3 (1.5)	
MRI and CT	134 (37.7)	54 (42.5)	13 (38.2)	67 (34.5)	
Intracranial vascular imaging					
MRA brain	323 (91.0)	116 (91.3)	30 (88.2)	177 (91.2)	0.76*
CTA brain	84 (23.7)	34 (26.8)	10 (29.4)	38 (19.6)	0.22
Conventional angiography, brain	51 (14.4)	28 (22.0)	8 (23.5)	15 (7.7)	0.0005
Cervical vascular imaging	187 (52.7)	63 (49.6)	22 (64.7)	103 (53.1)	0.29
MRA neck	154 (43.4)	49 (38.6)	17 (50.0)	88 (45.4)	0.35
CTA neck	45 (12.7)	17 (13.4)	7 (20.6)	21 (10.8)	0.28
Conventional angiography, neck	14 (3.9)	6 (4.7)	3 (8.8)	5 (2.6)	0.13*
Follow-up vascular imaging (any modality)					
At least one follow-up study	147 (41.4)	68 (53.5)	15 (44.1)	64 (33.0)	0.001
>1 follow-up study	68 (19.2)	39 (30.7)	6 (17.6)	23 (11.9)	<0.0001
Time to vascular imaging, days					
Stroke to baseline imaging, median (IQR)	1 (0, 2)	1 (0, 2)	1 (0, 2)	1 (0, 2)	0.81**
Stroke to 1st follow-up, median (IQR) (n=147)	79 (18, 128)	39 (12, 111)	70 (24, 105)	99 (44, 155)	0.002**
Stroke to last follow-up, median (IQR) (n=68)	277 (172, 408)	257 (106, 397)	212 (172, 355)	372 (219, 448)	0.15**

* Fisher's exact

**Kruskal-Wallis