Supplementary for Syntax

OVID Medline (PubMed) syntax

(moyamoya OR moya OR moya-moya [Title/Abstract]) AND (cognition OR neurocognitive OR intelligence OR psycho OR executive OR cognitive OR mental OR retardation OR memory OR language OR dementia [Title/Abstract])

Embase syntax

(moyamoya:ab,ti OR moya:ab,ti OR moya moya:ab,ti) AND (cognition:ab,ti OR neurocognitive:ab,ti OR intelligence:ab,ti OR psycho:ab,ti OR executive:ab,ti OR cognitive:ab,ti OR mental:ab,ti OR retardation:ab,ti OR memory:ab,ti OR language:ab,ti OR dementia:ab,ti)

Supplementary for the Risk Assessment

Newcastle–Ottawa Scale adapted[†] for cross-sectional studies

Selection: (Maximum 4 stars)

1) Representativeness of the sample[‡]
   a) Truly representative of the average in the target population*
   b) Somewhat representative of the average in the target population*
   c) No description of the derivation of the cohort

2) Sample size[§]
   a) Justified and satisfactory*
   b) Not justified

3) Selection criteria
   a) Selection criteria were clearly described and consecutive patients were included*
   b) Selection criteria were not clearly described and it was unclear whether consecutive patients were included

4) Ascertainment of the exposure[||]
   a) Validated measurement tool*
   b) Non-validated diagnostic measures (but the tool is available or described), or not all patients were DSA proven*
   c) No description of the diagnostic tool

Outcome: (Maximum 3 stars)

1) Assessment of the outcome (description of cognitive tests applied)[¶]
   a) Extensive neuropsychological evaluation**
   b) IQ*
   c) Screening test*
   d) No description

2) Quantitative data:
   a) The study reported cognitive or intellectual functioning in children and adults that allowed analysis of quantitative data.*
   b) The study did not report cognitive or intellectual functioning in children and adults that allowed analysis of quantitative data.

DSA, digital subtraction angiography; IQ, intelligence quotient.
The asterisk refers to the the number of stars (* or **) that can be assigned. It's a scoring method but not an actual footnote; [†]This scale has been adapted by the authors from the Newcastle-Ottawa Quality Assessment Scale for cohort studies[1] and the scale developed by Herzog et al. (2013)[2] to perform a quality assessment of cross-sectional studies for the systematic review: 'Cognitive functions in children and adults with moyamoya vasculopathy: a systematic review and meta-analysis.' Since there were no groups to compare (only patients with moyamoya (no control groups) were reviewed for this systematic review), we could not include the section 'Comparability'; [‡]Patients with moyamoya disease or syndrome: 1 star; [§]Sample size of n≥30: 1 star; [||] DSA or magnetic resonance angiography: 1 star; [¶]Neuropsychological test battery applied: 2 stars, IQ or screeningtest: 1 star.

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Supplementary Table 1. Applied cognitive instruments/tests for each study

Study	Applied instruments/tests*
Hsu et al. (2014)[3†]	WISC-III or WISC-IV; WAIS-III POI: Perceptual Organization Index WMI: Working Memory Index PSI: Processing Speed Index WL1: Immediate Recall of the Word List WL2: Delayed Recall of the Word List WL-recog: Recognition of the Word List CFT: Category Fluency Test JLO: Judgment of Line Orientation
Williams et al. (2012)[4†]	WISC-III or WISC-IV; WAIS-III; WPPSI-III VCI: Verbal Comprehension Index PRI: Perceptual Reasoning Index WMI PSI
Lee et al. (2011)[5†]	KEDI-WISC-R BGT recall: Bender Gestalt Test
Imaizumi et al. (1999)[6†]	WPPSI; WISC-R; WAIS-R; Tanaka-Bonet Intelligence Test Tumori-Inage Mental Development Test
Ohtaki et al. (1998)[7†]	WAIS-R; WISC-R
Matsushima et al. (1997)[8†]	WISC
Matsushima et al. (1991)[9†]	WISC; development questionnaires of Tsumori et al.
Sato et al. (1990)[10†]	WISC-R; WIPPSI; Developmental test BGT
Tagawa et al. (1989)[11†]	WISC
Ibayashi et al. (1985)[12†]	WAIS; Benton's Visual Memory Test
Ishii et al. (1984)[13†]	WISC; WAIS
Lei et al. (2017)[14‡]	TMT-B (s): Time consumed in the Trail Making Test part B MES-EX: executive subtests of Memory and Executive Screening
Kazumata et al. (2015)[15‡]	WAIS-III WSCT: Wisconsin Sorting Test TMT-A/B: Trail Making Test part A and B CPT: Continuous Performance Test Stroop test RST: Reading Span Test
Su et al. (2013)[16‡]	MoCA: Montreal Cognitive Assessment
Calviere et al. (2012)[17‡]	Letter R Category (animals) fluency test TMT-A/B Stroop interference condition Brixton test WCST-C/-P: Wisconsin Card Sorting Test number of categories and number of perseverations Colored dots and word sections of the Stroop test Verbal fluency tests Naming and Recognition Test of 80 common objects Rey figure copy test Hooper test Immediate and delayed 16 free and cued recalls Rey figure recall

Supplementary Table 1. Continued

Study	Applied instruments/tests*
Festa et al. (2010)[18‡]	WAIS-III; WASI Hopkins Verbal Learning Test California Verbal Learning Test TMT-A/B Boston Naming Test Animal Fluency COWAT: Controlled Oral Word Association Test WCST: Wisconsin Card Sorting Test Grooved Pegboard Test Hand Dynometer
Karzmark et al. (2008)[19‡]	WAIS-R; WAIS-III California Verbal Learning Test-II Memory Test-Revised Visual Reproduction subtest Delis-Kaplan Executive Function System Design Fluency Test FAS/AN: Letter and Category Fluency Tests TMT-A/B Grooved Pegboard Tactile Form Recognition Test Boston Naming Test

This table represents the cognitive instruments/tests used in each study separately.
WISC (-R or -III or -IV), Wechsler Intelligence Scale (revised or third or fourth edition); WAIS (-R or -III), Wechsler Adult Intelligence Scale (revised or third edition); WPPSI (-III), Wechsler Preschool and Primary Scale of Intelligence (third edition); KEDI-WISC-R, Korean Educational Development Institute Wechsler Intelligence Scale for Children-Revised; WASI, Wechsler Abbreviated Intelligence Scale.
*As reported by the authors; †Studies reporting results in children; ‡Studies reporting results in adults.

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Supplementary Table 2. Predefined cognitive domains according to standard neuropsychological practice specified in Lezak[20]

Cognitive domain	Included test
General intelligence	
Crystallised intelligence	Verbal IQ
	Similarities (WAIS)
	Vocabulary (WAIS)
	Information (WAIS)
	Comprehension (WAIS)
	National Adult Reading Test
	Synonyms
Fluid intelligence	Performal IQ
	Raven Progressive Matrices
	Picture Completion (WAIS)
	Picture Arrangement (WAIS)
	Arithmetic
	Category Test
Memory	
Working memory	Digit Span Forward & Backward
	Block Span Forward & Backward
	Memory Scanning Test
	Brown-Peterson task
Learning & Immediate memory	Logical Memory Immediate Recall
	Visual Reproductions Immediate Recall
	Paired Associate Learning Immediate Recall (verbal & nonverbal)
	Serial Digit Learning
	Word List Immediate Recall
	(Buschke) Selective Reminding Test Immediate Recall
	Visual Retention Test Immediate Recall
	Object Memory Immediate Recall
	Rey Complex Figure Immediate Recall
	Auditory Verbal Learning Test Immediate Recall
	Serial Learning Test
	Word/Picture Recognition Immediate Recall
	Spatial Memory Test
	California Verbal Learning Test Immediate Recall
	Claeson-Dahl Test Immediate Recall
	Seashore Tonal Memory Test
	Figural Memory Immediate Recall
	Iconic Memory
	Maze Learning Immediate Recall
	Tactual Performance Test Immediate
	Prose Recall Immediate Recall
	Symbol-Digit Learning Test
Learning & Immediate memory	Babcock paragraph Immediate Recall
	East Boston Memory Test Immediate Recall
Delayed memory	Logical Memory Delayed Recall
	Visual Reproductions Delayed Recall
	Word List Delayed Recall
	(Buschke) Selective Reminding Test Delayed Recall
	Visual Retention Test Delayed Recall
	Object Memory Delayed Recall
Cognitive domain	Included test
	Rey Complex Figure Delayed Recall
	Auditory Verbal Learning Test Delayed Recall
	Paired Associate Learning Delayed Recall (verbal & nonverbal)
	Word/Picture recognition delayed
	California Verbal Learning Test Delayed Recall
	Claeson-Dahl Test Delayed Recall
	Figural memory Delayed
	Maze Learning Delayed
	Tactual Performance Test Delayed Recall
	Delayed serial visual/verbal form memory task
	Prose Recall Delayed
	Babcock paragraph Delayed
	East Boston Memory Test Delayed Recall
	Logical Memory Delayed Recall
	Visual Reproductions Delayed Recall
	Word List Delayed Recall
	(Buschke) Selective Reminding Test Delayed Recall
	Visual Retention Test Delayed Recall
	Object Memory Delayed Recall
	Included test
	Rey Complex Figure Delayed Recall
	Auditory Verbal Learning Test Delayed Recall
	Paired Associate Learning Delayed Recall (verbal & nonverbal)
	Word/Picture recognition delayed
	California Verbal Learning Test Delayed Recall
	Claeson-Dahl Test Delayed Recall
	Figural memory Delayed
	Maze Learning Delayed
	Tactual Performance Test Delayed Recall
	Delayed serial visual/verbal form memory task
	Prose Recall Delayed
	Babcock paragraph Delayed
	East Boston Memory Test Delayed Recall

Supplementary Table 2. Continued

Cognitive domain	Included test
Processing speed	
Psychomotor efficiency	Digit Symbol Substitution
	Symbol Digit Modalities Test
	Trailmaking Test A
	Grooved Pegboard
	Purdue Pegboard
	Graded Reaction Time Task
	Perceptual Speed
Motor speed	Simple reaction time
	Fingertapping Test
	Finger Oscillation Test
Attention	
Visual attention	Stroop Color Word Test Part I & II
	Facial Recognition Test
	Target finding task
Sustained attention	Digit Vigilance Test
	Quatember & Maly's Vigilance Test
Divided attention	PASAT
Selective attention	Stroop Color Word Test Part III
Cognitive domain	Included test
Cognitive flexibility	Lexical Fluency Task
	Category Fluency Task
	Trailmaking Test B (also C, D and Color)
	Category Test
	Concept Shifting Task
	Wisconsin Card Sorting Task
	Serial subtraction (3s of 7s)
	Card Sorting
Perception & Construction	Visual Retention Test Copy
	Visual Reproductions Copy
	Block Design
	Clock Drawing
	Rey Complex Figure Copy
	Tactual Performance Test Part I
	Object Assembly (WAIS)
	Embedded Figures
	De Renzi Rods
	Flicker Fusion
	Perception of spaced stimuli
	Time judgement
	Visual Recognition Threshold
	Street Completion
	Rosen figure drawing test

Supplementary Table 2. Continued

Cognitive domain	Included test
Language	(Boston) Naming Test
	Token Test
	Boston Diagnostic Aphasia Test Writing Scale

IQ, intelligence quotient; WAIS, Wechsler Adult Intelligence Scale.

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Supplementary Table 3. Characteristics of studies assessing cognitive functions in children and adults with moyamoya vasculopathy

Study	Mid-year	Design	Inclusion criteria	Exclusion criteria	No.	Age (yr)	Female (%)	Ethnicity (%)	Presenting symptoms (%)	Duration (mo)	MMV site (%)	Site of stroke clinically (%)	Site of stroke imaging (%)
Hsu et al. (2014)[3]*	2010	-	Pediatric MMD >6 yr old; TIA as initial symptom	Cortical hemorrhage; prior revascularization; uncooperation; underlying systemic diseases	13	13.9±6.3 (6–17)†	-	Chinese	TIA 100	17±15.9 (1–48)†	-	-	-
Williams et al. (2012)[4]*	2004	Retro	MMD or MMS; <18 yr, NPA pre-surgery; English language skills	Whole brain radiation; severe developmental delay associated with genetic comorbidities; revascularization surgery; lack of parent/child agreement to NPA	30	10.1±4†	60	Caucasian 40 Asian 27 Black 20 Other 13	Infarction 50	35.0±49 (2–204)†	Bi 47 Uni 53	Bi 10 Uni 40	No stroke 30 Stroke 70 Bi 33 Uni 67 Cortical 57 WM 43
Lee et al. (2011)[5]*	2007	-	MMD with pre- and postoperative NPA	-	65	9.1 (4–17)†	43	Korean	-	-	Bi 82 Uni 18	-	No stroke 60 Stroke 40 MS 15 BZ 25
Imaizumi et al. (1999)[6]*	1984	-	MMD and IQ tested >once during course disease	-	38	6.5±3.3 (1–13)†	63	Japanese	Infarction 26 TIA 63 Other 11	16.2±16.1 (1–60)†	-	-	-
Ohtaki et al. (1998)[7]*	1990	Retro	Omental transplantation frontal lobes	-	8§	7.1±2.0 (5–11)‖	75	Japanese	Minor completed stroke 12.5 Hemorrhage 12.5 TIA 75	18.9±19.7 (2–60)‖	Bi 87 Uni 13	Bi 25 Uni 75	-
Matsushima et al. (1997)[8]*	-	Retro	IQ >70; EDAS performed <9.5 yr	-	20	9.6±3.4‖	40	Japanese	Infarction 30 TIA 70	-	-	-	-
Matsushima et al. (1991)[9]*	1984	-	MMD	-	50¶	9.4±4.3 (2–21)‖	56	Japanese	Movement disorder 80 Seizures 6 Headache 10 Involuntary movements 4	55.8±50.7 (0–188)‖	-	-	-
Sato et al. (1990)[10]*	-	-	Revascularization and CBF evaluation	-	12**	5.9±2.3 (1–10)†	33	Japanese	Ischemia 50 TIA 50	12.6±10.6 (1–31)†	Bi 92 Uni 8	Bi 66 Uni 33	No stroke 50 Stroke 50 Bi 50 Uni 50
Tagawa et al. (1989)[11]*	-	-	Children with MMD	-	10††	10.2±3.2 (6–15)†	60	Japanese	Infarction 10 TIA 90	57.8±50.5 (13–155)†	-	-	-
Ibayashi et al. (1985)[12]*	-	-	Juvenile MMD patients	-	15	9.2±3.3 (5–16)‡‡	53	Japanese	Completed stroke 53 TIA 47	48.3±44.3 (19–136)‡‡	-	Bi 73% Uni 27%	-
Ishii et al. (1984)[13]*	-	-	-	-	20	9.9±3.1 (5–16)‡‡	50	Japanese	Completed stroke 60 TIA 40	-	-	-	-

Supplementary Table 3. Continued

Study	Mid-year	Design	Inclusion criteria	Exclusion criteria	No.	Age (yr)	Female (%)	Ethnicity (%)	Presenting symptoms (%)	Duration (mo)	MMV site (%)	Site of stroke clinically (%)	Site of stroke imaging (%)
Lei et al. (2017)1[49§§]	2013	Pro	18–80 yr; rhanded; MMD on DSA; no abnormalities/ICH several brain locations; no surgery; physically able NPA	Significant neurological diseases; psychiatric disorders; other cerebrovasular diseases; systemic diseases; specific medication	26	40.2±9.4[††]	54	Chinese	Minor stroke 27 TIA 54 Headache 19	-	-	-	No hyperintense signals >8 mm in maximum dimension
Kazumata et al. (2015)[15§§]	2013	Pro	>20 yr; idiopathic MMD	Quasi MMD; cortical infarction/subcortical lesion >8 mm; intracranial hemorrhage; revasculari-zation surgery; neurological deficit because of stroke; comorbid illness affecting cognition	23	40.9±9.5 (21–58)[††]	74	Japanese	TIA 43 Asymptomatic 57	-	Bi 100	-	No stroke 57 Stroke 43 Bi 50 Uni 50
Su et al. (2013)[16§§]	2008	Pro	MMD with IVH; 18-60 yr; no revascularization surgery; BI >60/mRS <4; no mental disability	Other cerebrovascular diseases; AED; recurrent stroke during FU	26	43.7±8.6 (26–59)[††]	46	Chinese	Hemorhage 100	1.2[†]	-	-	IVH 100
Calviere et al. (2012)[17§§]	2002	Pro	MMD; >3 mo after stroke; no revascularization surgery	<18 yr; any associated disease potentially responsible for the arterial lesions	13	36.6±12.9[††]	64	Caucasian 86 Other 12	Ischemic stroke 62 Hemorrhage 8 Other 30	36.1[‖¶]	Bi 64 Uni 36	Bi 12 Uni 88	No stroke 29 Stroke 71 Bi 60 Uni 40 Cortical 70 SC 60 BZ 90 WM10
Festa et al. (2010)[18§§]	2002	Pro- and retro	MMD with complete NPA	(neurological) Disorders affecting cognition	29	39.9±11.2 (20–65)[‖¶]	62	Caucasian 59 Hispanic 20 Afro-american 20 Asian 21	Ischemic stroke 72 TIA 17 Hemorrhage 3 Other 8	-	Bi 86 Uni 14	-	No stroke 17 Stroke 83 Bi 75 Uni 25
Karzmark et al. (2008)[19§§]	2005	–	MMD	-	36	36.6±9.9[†]	67	Caucasian 75 Asian 17 Other 8	-	-	-	-	-

Values are presented as mean±standard deviation (range), mean±standard deviation, or mean (range). This table represents the study and patients' characteristics separated for children and adults.
MMV, moyamoya vasculopathy; MMD, moyamoya disease; TIA, transient ischemic attack; Retro, retrospective; MMS, moyamoya syndrome; NPA, neuropsychological assessment; Bi, bilateral; Uni, unilateral; WM, white matter; MS, major stroke; BZ, borderzone; IQ, intelligence quotient; EDAS, encephaloduroateriosynangiosis; CBF, cerebral blood flow; Pro, prospective; DSA, digital subtraction angiography; ICH, intracerebral hemorrhage; IVH, intraventricular hemorrhage; BI, Barthel Index; mRS, modified Rankin Score; AED, anti-epileptic drug; FU, follow-up; SC, subcortical; R, right.
*Studies reporting results in children; [†]At NPA; [‡]At diagnosis; [§]Excluding 2 patients (1 scaled out, 1 not investigated); [‖¶]At operation; [¶]Study included 65 patients with preoperative data in 50 patients; [**]Study included 13 patients from which 12 had preoperative data; [††]Study included 21 patients from which 10 had preoperative data; [††]Not specified; [§§]Studies reporting results in adults; [‖¶]At presentation.

Supplementary Table 4. Neuropsychological test performances

Study	Authors criteria cognitive impairment	Cognitive impairment overall (%)	Conclusion authors	Cognition screener score	% Impaired	(FS) IQ score	% Impaired	VIQ score	% Impaired	PIQ score	% Impaired	DQ score	% Impaired	Memory score	% Impaired	Procspeed score	% Impaired	Att/EF score	% Impaired	Visper/ const score	% Impaired	Language score	% Impaired
Hsu et al. (2014)[3]*	IQ: >80 normal 70–79 borderline <70 defective NPA: <1.5 SD borderline <2 SD defective	39	Normal intellectual development with specific impairments in some	-	-	102±13 (82–124)	0	99±15 (77–117)	17	103±13 (81–123)	0	-	-	(z=–0.39) 8.6±2.8	15	(z=0.00) 103.2±17.9	8	(z=0.50) 13.3±4.7	8	(z=–0.53) 21.8±4.3	18	-	-
Williams et al. (2012)[4]*	1 SD from the mean (IQ, 85–110)	-	Significant lower than test sample	-	-	87±18	-	91±14	-	89±22	-	-	-	(z=–0.85) 87.3±15.8	-	(z=–0.86) 87.2±13.2	-	-	-	-	-	-	-
Lee et al. (2011)[5]*	Compared with population averages	-	Age appropriate IQ	-	-	107±14	-	108±13	-	105±16	-	-	-	(z=0.45) 3.8±1.9	-	-	-	-	-	-	-	-	-
Imaizumi et al. (1999)[6]*	-	-	-	-	-	93±23	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Ohtaki et al. (1998)[7]*	IQ: >90 normal 89–70 borderline <69 retardation	13	Normal intellectual range	-	-	103±20 (58–128)	13	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Matsushima et al. (1997)[8]*	Normal IQ >86	15	-	-	-	107±18	-	105±21	–	109±13	-	-	-	-	-	-	-	-	-	-	-	-	-
Matsushima et al. (1991)[9]*	Normal IQ >86	50	-	-	-	-	-	-	-	-	-	84±30 (20–138)	50	-	-	-	-	-	-	-	-	-	-
Sato et al. (1990)[10]*	IQ: normal ≥1SD borderline –2SD to –SD: mild –3SD to –2SD moderate –3SD to –4SD	67	-	-	-	-	-	77±12[†] (58–88)	57	81±19[†] (42–104)	56	61±17[§] (42–72)	100	-	-	-	-	-	-	-	-	-	-
Tagawa et al. (1989)[11]*	-	30	Poor mental prognosis was correlated with early onset MMD	-	-	101±22 (71–134)	30	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Ibayashiet al. (1985)[12]*	-	-	IQ was reduced with advancing age	-	-	98±19	-	97±16	-	97±17	-	-	-	-	-	-	-	-	-	-	-	-	-
Ishii et al. (1984)[13]*	-	22	-	-	-	97±20	22	95±18	21	97±21	26	-	-	-	-	-	-	-	-	-	-	-	-
Lei et al. (2017)[14]‖	-	-	MMD patients performed worse than healthy controls	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Kazumata et al. (2015)[15]‖	-	30	MMD impairs executive funtion,working memory and attention	-	-	94±13	8	95±13	4	93±11	17	-	-	-	35	-	33	-	30	-	22	-	39

Supplementary Table 4. Continued

Study	Authors criteria cognitive impairment	Cognitive impairment overall (%)	Conclusion authors	Cognition screener score	% Impaired	(FS)IQ score	% Impaired	VIQ score	% Impaired	PIQ score	% Impaired	DQ score	% Impaired	Memory score	% Impaired	Procspeed score	% Impaired	Att/EF score	% Impaired	Visper/ const score	% Impaired	Language score	% Impaired
Su et al. (2013)[16]‖	Abnormal: MoCA <25 MCI: MoCA <25≥14	0	No impairment	27.4±1.2	0	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Calviere et al. (2012)[17]‖	Impairment: z-score ≥1.7SD below normative mean EDS: impairment ≥3 tests	54	-	-	-	-	-	-	-	-	-	-	-	(z=–0.4)	54	(z=–1.7)	23	(z=–0.95)	54	(z=–0.5)	23	(z=–0.15)	31
Festa et al. (2010)[18]‖	Z-score ≥2 domains >1.5SD or ≥1 domain >2SD below normative mean	69	Disruption in a broad range of functions	-	-	99±17	-	-	-	-	-	-	-	z-score –1.1±1.4	39	z-score –0.8±1.1	21¶	z-score –0.4±0.8	19**	z-score –0.4±1.3	29	z-score –0.8±1.1	20
Karzmark et al. (2008)[19]‖	>50% of the scores ≥1–2SDs below the mean	31	MMD can affect cognition (mostly EF)	-	-	95±9 (z=–0.6)	19	93±8 (z=–0.5)	25	93±8 (z=–0.5)	25	-	-	(z=–0.2)	7	(z=–0.9)	39	(z=–0.9)	43	(z=–0.2)	23	(z=–0.6)	40

Values are presented as mean±standard deviation (range) or mean±standard deviation. This table is divided into overall cognitive results of the studies separated for children and adults, followed by the test results for the cognitive screener test and all the six cognitive domains.

(FS)IQ, (full-scale) intelligent quotient; VIQ, verbal intelligence quotient; PIQ, performal intelligence quotient; DQ, developmental quotient; Procspeed, processing speed; Att, attention; EF, executive function; Visper/const, visual perception/construction; IQ, intelligence quotient; NPA, neuropsychological assessment; SD, standard deviation; MMD, moyamoya disease; MoCA, Montreal Cognitive Assessment; MCI, mild cognitive impairment; EDS, executive dysfunction syndrome.

*Studies reporting results in children; †n=7; ‡n=9; §n=3; ‖Studies reporting results in adults; ¶n=19; **n=16.

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Supplementary Table 5. Longitudinal neuropsychological test performances

Study	FU period (mo)	Surgery type (n)	Impairment overall (%), A/B	Conclusion authors, A/B	% Improved	% Stable	% Deteriorated	Cognition screener score, A/B	% Impaired A/B	(FS)IQ score, A/B	% Impaired A/B	VIQ score, A/B	% Impaired A/B	PIQ score, A/B	% Impaired A/B	DQ score, A/B	% Impaired A/B	Memory score, A/B	% Impaired A/B
Lee et al. (2011)[5]*	19† (5–46)	ID 65 Bifr 42	-	Functions are maintained well before and after surgery	-	-	-	-	-	107±14/ 108±13	-	108±13/ 106±13	-	105±16/ 109±31	-	-	-	(z=0.45) 3.8±1.9/ (z=0.77) 4.5±1.7	-
Imaizumi et al. (1999)[6]*	>120†	C 5 ID 13	-	No improvement	-	-	-	-	-	93±23/-	-	-	-	-	-	-	-	-	-
Ohtaki et al. (1998)[7]*	85.2±32.59§ (23–110)	C+Bifr 8	13/13	Stable	12	63	25	-	-	103±20 (58–128)/ 96±25 (48–138)	13/13	-	-	-	-	-	-	-	-
Matsushima et al. (1997)[8]*	113†	ID 20	15/20	-	-	-	-	-	-	107±18/ 100±16	–	105±21/ 100±16	–	109±13/ 100±16	-	-	-	-	-
Matsushima et al. (1991)[9]*	26.2±14.7† (7–58)	ID 41‖	50/49	Stable	27	49	24	-	-	-	-	-	-	-	-	84±30 (20–138)/ 83±32 (35–140)	50/ 49	-	-
Sato et al. (1990)[10]*	44.4±26.3¶ (4–99)	D 1 C 1 ID 10	67/58	-	PIQ 11 VIQ 29 DQ 0	PIQ 78 VIQ 57 DQ 100	PIQ 11 VIQ 14 DQ 0	-	-	-	-	77±12 (58–88)/ 82±25 (43–112)	57/29	81±19 (42–104)/ 79±24 (41–113)	56/56	61±17 (42–72)/ 56±10 (45–62)	100/ 100	-	-
Ibayashi et al. (1985)[12]*	6.5±4.9† (1–17)	C 2 ID 13	-	Surgery is considered to be effective	FSIQ 47 VIQ 20 PIQ 60	-	-	-	-	98±19/ 99±20	-	97±16/ 94±16	-	97±17/ 102±18	-	-	-	-	-
Ishii et al. (1984)[13]*	6–68†	C 2 ID 18**	22/-	Improved	FSIQ 53 VIQ 13 PIQ 67	FSIQ 40 VIQ 73 PIQ20	FSIQ 6 VIQ 13 PIQ 13	-	-	97±20/-	-	95±18/-	-	97±21/-	-	-	-	-	-
Su et al. (2013)[16]††	24§	-	0/100	Deteriorated	0	0	100	27.4±1.2/ 18.7±1.3	0/100	-	-	-	-	-	-	-	-	-	-

Values are presented as median (range), mean±standard deviation, mean±standard deviation (range), mean±standard deviation, or range. This table is divided into overall cognitive results at follow-up of the studies separated for children and adults, followed by the test results for the cognitive screener test and the available cognitive domains.

FU, follow-up; A, prior neuropsychological test result; B, longitudinal neuropsychological test result; (FS)IQ, (full-scale) intelligent quotient; VIQ, verbal intelligence quotient; PIQ, performal intelligence quotient; DQ, developmental quotient; ID, indirect; Bifr, bifrontal; C, combined; D, direct.

*Studies reporting results in children; †FU period defined as time of operation to NPA; ‡FU period defined time from onset of disease to NPA; §FU period defined as time of NPA to NPA; ‖41 out of the 50 patients investigated postoperatively; ¶FU period unspecified; **15 out of the 20 patients investigated postoperatively; ††Studies reporting results in adults.

Supplementary Table 6. Critical appraisal of the included studies

Study	Study design	Selection				Outcome	
		Representativeness of the sample	Sample size	Selection criteria	Ascertainment of exposure	Assessment outcome	Quantitative data
Hsu et al. (2014)[3]*	Cross-sectional	+		+	+	++	+
Williams et al. (2012)[4]*	Cross-sectional	+	+	+	+	++	+
Lee et al. (2011)[5]*	Cross-sectional	+	+	+	+	++	+
Imaizumi et al. (1999)[6]*	Cross-sectional	+	+	+		+	+
Ohtaki et al. (1998)[7]*	Cross-sectional	+		+	+	+	+
Matsushima et al. (1997)[8]*	Cross-sectional	+		+	+	+	+
Matsushima et al. (1991)[9]*	Cross-sectional	+	+	+		+	+
Sato et al. (1990)[10]*	Cross-sectional	+		+	+	+	+
Tagawa et al. (1989)[11]*	Cross-sectional	+		?†	?†	+	+
Ibayashi et al. (1985)[12]*	Cross-sectional	+		?†	?†	+	+
Ishii et al. (1984)[13]*	Cross-sectional	+				+	+
Lei et al. (2017)[14]‡	Cross-sectional	+	+	+	+	+	+
Kazumata et al. (2015)[15]‡	Cross-sectional	+		+	+	++	+
Su et al. (2013)[16]‡	Cross-sectional	+		+	+	+	+
Calviere et al. (2012)[17]‡	Cross-sectional	+		+	+	++	+
Festa et al. (2010)[18]‡	Cross-sectional	+		+	+	++	+
Karzmark et al. (2008)[19]‡	Cross-sectional	+	+		+	++	+

*Studies reporting results in children; †This information could not be extracted by our translators; ‡Studies reporting results in adults.

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Supplementary Table 7. Linear regression analysis

Authors	Cognitive impairment overall (%)	Mean age	Duration symptoms (mo)	% Female	% Infarction	% TIA(s)
B (95 CI; *P*)		–0.014 (–0.112 to 0.083; 0.723)	0.000 (–0.016 to 0.016; 0.945)	–0.005 (–0.025 to 0.014; 0.508)	–0.002 (–0.017 to 0.013; 0.672)	–0.002 (–0.013 to 0.017; 0.672)
Hsu et al. (2014)[3]*	39	13.9±6.3 (6–17)	17±15.9 (1–48)	-	0	100
Ohtaki et al. (1998)[7]*	13	7.1±2.0 (5–11)	18.9±19.7 (2–60)	75	-	-
Matsushima et al. (1997)[8]*	15	9.6±3.4	–	40	30	70
Matsushima et al. (1991)[9]*	50	9.4±4.3 (1.6–21)	55.8±50.7 (0–188.4)	56	-	-
Sato et al. (1990)[10]*	67	5.9±2.3 (1–10)	12.6±10.6 (1–31)	33	31	69
Tagawa et al. (1989)[11]*	30	10.2±3.2 (6–16)	57.8±50.5 (13–155)	60	10	90
Ishii et al. (1984)[13]*	22	9.9±3.1 (5–16)	-	50	60	40
B (95 CI; *P*)		–0.044 (–0.184 to 0.096; 0.387)	-	0.011 (–0.031 to 0.053; 0.460)	-	-
Kazumata et al. (2015)[15]†	30	40.9±9.5 (21–58)	-	74	-	-
Su et al. (2013)[16]†	0	43.7±8.6 (26-59)	-	46	-	-
Calviere et al. (2012)[17]†	54	36.6±12.9	-	64	-	-
Festa et al. (2010)[18]†	69	39.9±11.2 (20–65)	-	62	-	-
Karzmark et al. (2008)[19]†	31	36.6±9.9	-	67	-	-

Values are presented as mean±standard deviation (range) or mean±standard deviation. This table represents the results of the linear regression analysis weighed by the inverse standard error of the proportion of patients with impaired cognition for the available patients' characteristics.
TIA, transient ischemic attack; CI, confidence interval.
*Studies reporting results in children; †Studies reporting results in adults.

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