Surgical Treatment of Paediatric Moyamoya Disease: A Single Centre Review

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Background and aim

- Moyamoya disease (MMD): progressive stenosis of distal internal carotid arteries (ICA)
- Classically, definitive treatment in the paediatric group includes 1) indirect revascularisation, 2) direct—STA-MCA bypass and 3) combined direct and indirect approach [1]
- At our centre, practice has been changing over the past 5 years. With increasing maturity in bypass technique which allows for safer operation, combining direct bypass with indirect revascularisation is now preferred in treating paediatric MMD
- Aim of this study: Review the outcomes of MMD revascularisation procedures performed at our centre from 2000 to 2019

Method

- Retrospective descriptive review
- All paediatric MMD patients who have received treatment at our hospital from 2000 to 2019 have been included
- Data retrieved included demographics, type of operations, surgical and long-term outcomes

Results

12 patients included, 19 operations, 20 hemispheres operated on

Diagram 1: Number of revascularisation procedures (19 in total)

Table 1: Patients characteristics

	Combined Direct and Indirect (n=5)	Indirect revascularisation (n=7)
Number of patients Male Female	 2 3	 4 3
Median age of first operation	10 yr old (youngest: 3yr old)	8 years old (youngest: 3 mth old)
Median follow-up time	29 months	81 months
Bypass vessel characteristics	Mean vessel caliber: 0.9mm	--

1. Operative outcomes

Diagram 2: Mean operative blood loss

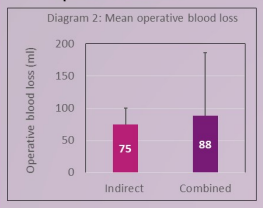
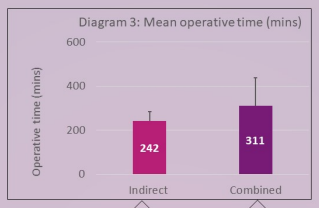


Diagram 3: Mean operative time (mins)



DSA not performed

DSA + direct revascularisation

Diagram 4: Combine Direct Bypass and Indirect Revascularisation — DSA intra-op in hybrid theatre to save one GA session

Operative blood loss and the operative time in the combined approach is reasonable as compared to those of the indirect approach

2. Clinical and radiological outcomes

Measurement of Outcome	Combined (n = 5)	Indirect revascularisation (n = 7)
Symptom improvement	5 (100%)	7 (100%)
Improved perfusion on imaging	5 (100%)	7 (100%)
Mean duration between operation and clinical improvement	48 days	77 days
Residual neurological deficit at last follow-up	0	2

Table 2: Clinical and Radiological Outcomes

3. Complications

Combined Group

- 1 new infarct
- Documented suboptimal perioperative blood pressure control
- Neurological deficit subsequently resolved

Indirect Group

- 1 new infarct
- Neurological deficit subsequently resolved

Combined direct bypass and indirect revascularisation allows faster symptom improvement and better long-term neurological outcome

Case sharing

Patient A

- Male, now 6 years old
- Presented at 3 years old with left hemiparesis
- Angiogram showed truncation of Right M1
- Combined direct and indirect revascularisation operation was performed in 2016 (3 years old)
- The patient demonstrated significant symptom improvement with complete resolution of hemiparesis within 1 month post-op

Diagram 5a: Pre-op MR angiogram

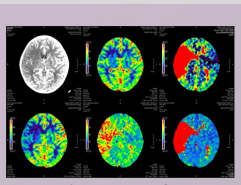


Diagram 5b: Pre-op CT perfusion scan

Direct STA-MCA bypass

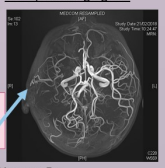


Diagram 5c: MR angiogram 2-years post-op

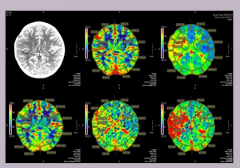


Diagram 5d: CT perfusion scan 3-years post-op

Discussion

- Indirect revascularisation has been advocated as the primary treatment for paediatric Moyamoya disease in many centers because small vessel caliber creates technical challenges in direct bypasses
- Our centre has been performing combined direct and indirect revascularisation operation in paediatrics with reasonable operative time and blood loss as shown in our results
- Candidate selection: Risk of clamping and chance of producing ischaemic insult is particularly high in a "hungry brain"
- Good collaboration with Anesthetist → ensure stable blood pressure control and avoid hypocapnia

Conclusion

- Combined direct-indirect revascularisation: 1) Favorable surgical outcomes, 2) reasonable complication, 3) faster symptom resolution, 4) better long term neurological outcome
- Combined direct-indirect revascularisation should be the preferred approach in the paediatric population

Reference

Jay R, Craig T, Orlando S, Suresh M. Surgical treatment of pediatric Moyamoya disease. J Pediatr Neuroradiol 2014; 3(1): 29-37