



## Introduction

- The natural history of hemorrhagic (Figure 1) Moyamoya disease (MMD) is poor due to the high rehemorrhage rates reported to be between 32-61%.
- Post-revascularization the rehemorrhage rates have been reported to decrease to 12-17%.
- We aimed to establish the long term functional outcomes and rehemorrhage rates of hemorrhagic MMD patients treated with surgical revascularization and examine it in relation to the clinical and radiological factors.

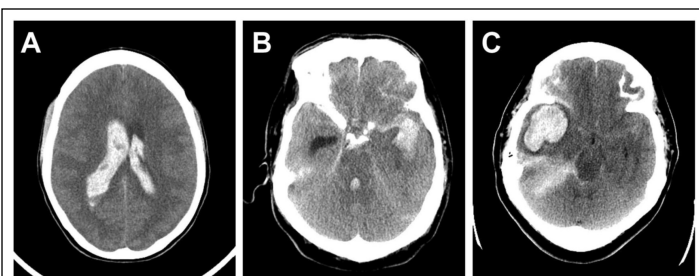


Figure 1: Different types of haemorrhage pattern in Moyamoya disease including intraventricular haemorrhage (A); intracerebral and subarachnoid haemorrhage (B) and intracerebral haemorrhage (predominant pattern, C)

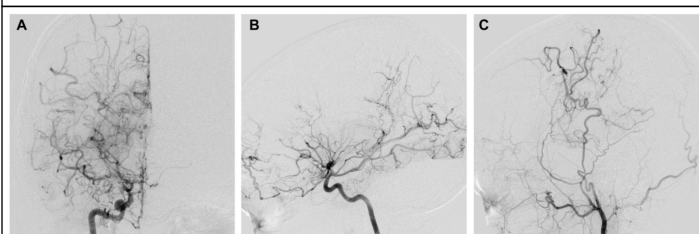


Figure 2: Cerebral DSA (A) AP and (B) lateral shows advanced stage Moyamoya disease involving right supraclinoid ICA, proximal MCA and ACA. (C) Post direct revascularization external carotid artery injection (lateral view) demonstrates perfusion of the MCA territory via the bypass.

## Methods

- All patients treated surgically for hemorrhagic MMD (Figure 2) over 23 years were identified from a prospective database.
- Modified Rankin scale (mRS) was used to grade the clinical status at presentation and assess outcomes with good being defined as mRS 0-2.
- Multivariate logistic regression analysis was performed after performing a stepwise selection process to determine the risk factors influencing the rehemorrhage rates and functional outcomes.

## Results

- 104 patients (69 female; 35 male) with a mean age of 39.3 years (3-64) were included.
- 81 patients had bilateral MMD while 23 had unilateral MMD.
- The mean mRS score (Table 1) at baseline was 2.31 (0-5).
- Of 172 revascularized hemispheres, 157 were direct superficial temporal artery (STA) - middle cerebral artery (MCA) bypasses; 15 were indirect.
- The mean follow-up duration was 61.44 months (0.25 - 239.8 months).
- Over the follow-up period, eight patients (7.7%) experienced rehemorrhage resulting in the rehemorrhage rate per person per year being 1.9%. Four patients died with one related to the rehemorrhage. At the last follow-up, the mean mRS score (Table 1) improved to 2.11.
- No significant risk factors were identified in relation to the rehemorrhage rates.
- The patients' initial mRS score was positively associated with their performance status at the final follow-up ( $p < 0.001$ ).
- Direct STA-MCA bypass (including those carried out in combination with an indirect procedure on the other hemisphere) was associated with better performance status (lower eventual mRS scores;  $p = 0.033$ ).

mRS score	Number of patients preoperatively	Number of patients at the last follow-up
0	35	54
1	34	21
2	14	13
3	11	6
4	9	4
5	1	2
6	0	4

**TABLE 1. Number of hemorrhagic Moyamoya disease patients per modified Rankin scale score preoperatively and at the last follow-up postoperatively**

## Conclusions

- Surgical revascularization of hemorrhagic MMD patients reduced the rehemorrhage rates to 7.7% (1.9%/person/year) compared with much higher historical natural history rates and led to an improvement in patients' performance status.
- The reported outcomes support the strategy to perform revascularization procedure in these patients with a preference for direct STA-MCA bypasses.

## Learning Objectives

The participant should be able to a) demonstrate awareness of the current literature on hemorrhagic MMD and its natural history; b) appreciate the controversies in the management of this disease entity and, c) in light of the presented results understand the rationale for performing surgical revascularization in this patient group.

## References

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