

## Introduction

Moyamoya disease is a vascular disorder characterized by progressive stenosis of the internal carotid artery. Clinical presentation often results from cerebral ischemia due to reduced perfusion within the cerebral hemispheres or hemorrhaging from the newly-formed collateral arteries. In the North American population, studies have reported a unimodal age distribution that peaks during the third, fourth, or fifth decades. However, the presentation, progression, treatment options, and post-operative clinical outcomes for elderly Moyamoya patients have never been reported.

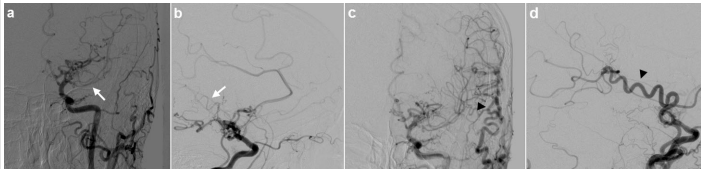
## Results

Seventy patients were diagnosed with probable or definite Moyamoya disease during the study period. Eight patients (6 females: 2 males; median age 63.5; range 62-71 years) were found to be sixty years or older and were included in the study. All patients had a Modified Rankin Scale (mRS) of either one or two (median 1) pre-operatively. Six patients (75%) underwent surgical treatment on a total of 7 hemispheres. Post-surgery, one patient had an improved mRS score, and four had no changes in their mRS scores. One patient was lost to follow-up. Of note, both patients who did not undergo surgical interventions suffered from intraparenchymal hemorrhages post-diagnosis.

## Methods

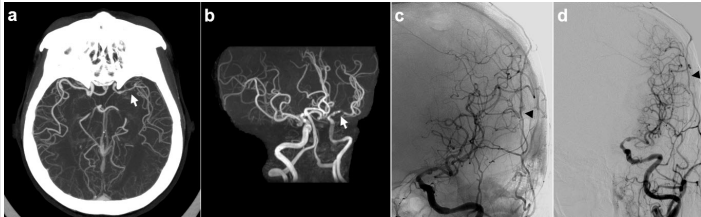
A retrospective analysis of all patients who were diagnosed with Moyamoya disease by the senior authors between 1991 and 2016, was performed. Patients who were 60 years or older at the time of treatment were further evaluated.

### Figure 1



Preoperative digital subtraction angiography (DSA) coronal (A) and sagittal (B) imaging showed near complete occlusion of the left middle cerebral artery (white arrows) along with moderate lenticulostriate collateral formation. Post STA-MCA (EC-IC) bypass, collaterals were seen emanating from the STA (black arrows) on coronal (C) and sagittal images (D). Revascularization was deemed consistent with Matsushima Grade C.

## Figure 2



Computed tomography angiography (CTA; A) and magnetic resonance angiography (MRA; B) indicate significant stenosis of the middle cerebral artery (white arrows), leading to dramatically reduced perfusion of the cerebral hemisphere. Placement of the superficial temporal artery (black arrows) on the top of the cortex, via an EDAS procedure, prompted significant neovascularization. Collateralization and revascularization are consistent with Matsushima Grade B.

### Table 1

Case	Age, Sex	Age at Time of Surgery	Ethnicity	Relevant Family History	Presentation	Elapsed Time Between Initial Symptomatic and Diagnosis (Yr.)	Findings on Pre-op Imaging	Type (Definite, Probable)	Maymura Grade	Surgical Procedure (IDR, Bypass)	Findings on Post-op Imaging
1	48, M	47	Caucasian	Stroke	CVA (Ischemic); Neurological deficits	5	IRM	Definite	Suzuki Stage V (Bilaterally)	Direct (L)	N/A
2	62, F	NS	African American	CAD	CVA (Hemorrhagic); Neurological deficits	7	IRM	Definite	Suzuki Stage V (L); Suzuki Stage IV (R)	NS	N/A
3	62, F	60	Caucasian	CAD	Ischemic; Neurological deficits	1	IRM	Definite	Suzuki Stage V (Bilaterally)	EDAS (L)	Matsumura Grade II
4	62, M	57	Caucasian	CAD	CVA (Hemorrhagic); Neurological deficits; Headaches	Concurrent	UM	Probable	Suzuki Stage IV (L)	Direct (L)	Matsumura Grade II
5	68, F	66	Asian	Noncontributory	Neurological deficits; Headaches	2	UM	Probable	Suzuki Stage IV (L)	EDAS (L)	Matsumura Grade C
6	71, F	NS	Caucasian	Stroke	Neurological deficits; Bitemporal atrophy	<1	UM	Probable	Suzuki Stage IV (L)	NS	N/A
7	63, F	60	Caucasian	Noncontributory	Ischemia (transient); Headaches	Unclear	IRM	Definite	Suzuki Stage III (Bilaterally)	EDAS (Bilaterally)	Matsumura Grade C; New infarcts near L ACA and bilateral MCA
8	64, F	61	Caucasian	Noncontributory	Ischemia	Concurrent	UM	Probable	Suzuki Stage III (R)	EDAS (R)	Matsumura Grade C

## Case Series for Elderly Moyamoya Patients

## Table 2

Case	Age, Sex	Elapsed Time Between Surgery and Clinical Follow up (months)	Initial mRS	Final mRS	Change in mRS	Progressive Symptomology	Post-Op Progressive Symptomology
1	68, M	N/A	1	1	0	Yes	N/A
2	62, F	NS	2	2	0	Yes	Yes
3	62, F	23	2	1	-1	Yes	No
4	62, M	17	2	2	0	Yes	No
5	68, F	6	1	1	0	Yes	No
6	71, F	NS	1	1	0	Yes	Yes
7	63, F	3	1	4	+3	Yes	Yes
8	64, F	37	1	1	0	No	No

## Post-Operative Changes in Functional Independence and Disease Progression

## Conclusions

Moyamoya disease is most commonly seen in young and middle-aged patients. Presentation in the elderly (defined as sixty years and above in this study) is rare, and has never been reported in literature. In this study, both direct and indirect revascularization procedures demonstrated potential benefit in treatment of these patients, with stabilization of progressive symptoms.