

Neurorehabilitation Outcome in Moyamoya Disease

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Moyamoya is a disease characterized by occlusion of the internal carotid, anterior, and middle cerebral arteries with associated rich collateral flow that presents a cloudy appearance on angiogram resembling a puff of smoke. The disease is most often progressive with associated hemiparesis and cognitive impairment. The functional outcome of patients with moyamoya is not well described in the literature. We describe four women (ages 25-36) who were transferred to a rehabilitation service after an average 17 days (12-26 days) in an acute care setting. Initial functional impairment was estimated using the Functional Independence Measure (FIM) score after discharge from inpatient rehabilitation (23-53 days) and was compared to the Uniform Data System for Medical Rehabilitation (UDSMR) for "first stroke" patients. Average admission FIM scores were similar in the two groups. The patients with moyamoya had a higher discharge FIM, longer length of stay, and slower rate of progress. Data on long-term survival and functional level would be useful, but it appears patients with moyamoya disease may benefit from rehabilitation oriented toward neurological deficits.

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TAKEUCHI AND SHIMIZU¹ originally described the case of a 29-year-old man with bilateral dysplasia of the internal carotid arteries. Angiography revealed an abnormal configuration of the vessels that appeared to resemble a "puff of smoke" or moyamoya in Japanese. Subsequently, various labels other than moyamoya were used to describe this vascular disorder, including obstruction of the circle of Willis, basal-normal vascular network, juxtabasal telangiectasia, cerebral arterial rete, rete mirabile, and Nishimoto's disease.²

Guidelines for diagnosis vary, but stenoses in the supraclinoid internal carotid arteries (ICA) bilaterally and in the anterior and middle cerebral arteries (ACA and MCA, respectively), as well as the moyamoya phenomenon bilaterally with no underlying disease, have been described.^{2,3}

The etiology is unknown, but the two main theories about the development of moyamoya is that it is either congenital or acquired. The two may not be completely incompatible, and many factors, including predisposition, systemic infection, focal

factors, and platelet abnormalities, appear to contribute to the development of the disease.

There is limited information regarding outcome or functional capacity following medical or surgical treatment.⁴⁻⁹ In addition, no studies have focused on rehabilitation outcome of moyamoya patients. These patients may evidence significant motor, sensory, cognitive-linguistic, and functional deficits, but because the disorder is relatively infrequent, it is difficult to assemble large groups for study. This article reports four cases of verified moyamoya disease and neurorehabilitation outcome and describes the similarities and differences with the typical stroke.

CASE REPORTS

Medical data, laboratory tests, radiological findings, and Functional Independence Measure (FIM) scores for the four patients are presented in table 1. The FIM is a standardized measure of functional skills.¹⁰ Functional skills are rated on a numerical scale from 1 (total assistance) to 7 (complete independence). Patient scores on individual activities are summed to achieve various functional skill area scores. These areas and maximum scores include self care (42), sphincter control (14), mobility (21), locomotion (14), communication (14) and social cognition (21).¹⁰ An occupational therapist, physical therapist, and speech language pathologist were involved with the rehabilitation program of all patients. Conventional methods of stroke rehabilitation were used and included combinations of range of motion and muscle strengthening exercises, mobilization activities, and compensatory techniques. No specific neurophysiologic therapy approach was used (eg, Bobath), nor was electrical stimulation used in any of the four patients.

Case 1

The patient was a 25-year-old woman with a history of tubal ligation and hypothyroidism who had an abortion 2 weeks before presenting with an acute headache and left hemiparesis. Current medications included levothyroxine. Computed tomography (CT) showed acute right basal ganglia hemorrhage. Angiography revealed severe right ACA and MCA stenosis and abnormal netlike vessels at the base of the brain consistent with moyamoya (fig 1). The patient remained in acute care for 12 days.

Physical examination upon admission to rehabilitation revealed an obese woman with sensation decreased on the left side of her face, arm, and leg. There were no sensory deficits on the right. Motor strength in the left upper limb was: 3/5 deltoid, triceps, 2/5 biceps, 1/5 wrist extension and flexion, and 2/5 for grip. Left lower extremity strength was 4/5 except for dorsiflexion at the ankle, which was 1/5. Right sided muscle strength was 5/5 throughout. Muscle stretch reflexes were increased throughout the left and normal on the right, and no clonus was present. Speech was dysarthric secondary to muscle weakness and incoordination. The patient evidenced deficits in all areas of functional skills except for communication and cognition. Her most prominent impairment was in locomotion (scale score = 2) and secondarily in mobility (scale score = 11) and self care (scale score = 21). By discharge, her speech, left sided strength, and fine motor control had improved markedly. She was ambulating on all terrains without the use of assistive devices. After 30 days in rehabilitation, she evidenced

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Table 1: Summary of Cases

Case	Age/Race/Sex	Medical History	Presenting Symptoms	Radiologic Findings	FIM		
					ADM	D/C	FU
1	25/AA/F	Tubal Ligation Hypothyroidism Abortion	Acute headache Left hemiparesis	Right basal ganglia hemorrhage Right ACA/MCA stenosis	78	113*	119 (6 mos)
2	35/C/F	Hypertension Otitis Media Hypothyroidism	Loss of consciousness Atypical migraines	Ventriculomegaly Bilateral supraclinoid stenosis ICA Left choroidal aneurysm and ICH	83	112*	114 (6 mos)
3	28/C/F	Hypertension Hypercholesterolemia Anton's syndrome Major depression	Left hemiparesis Dysarthria Blindness	Bilateral supraclinoid ICA/MCA stenosis Bilateral occipital infarcts Cortical atrophy	36	60*	60 (12 mos)
4	36/C/F	Hypertension Ruptured ovarian cyst Hypoalbuminemia	Dysarthria Right Hemiparesis "Depression"	Bilateral supraclinoid ICA stenosis Left MCA infarct	51	86*	93 (5 mos)

Abbreviations: AA, African-American; C, Caucasian; ADM, admission; D/C, discharge; FU, follow-up; F, female; ACA, anterior cerebral artery; MCA, middle cerebral artery; ICA, internal carotid artery; ICH, intracerebral hemorrhage.

* $p < .001$.

improvement in all areas of functioning (table 1), and at follow-up she was functioning maximally in all skills areas except for self care (scale score = 36).

Case 2

The patient was a 35-year-old woman with a history of thyroidectomy and associated hypothyroidism requiring pharmacologic replacement, mild hypertension, headaches, and recurrent left otitis media. She presented with "atypical migraines" and a loss of consciousness. CT showed intraventricular hemorrhage and ventriculomegaly; therefore, a ventriculostomy was performed. An arteriogram revealed bilateral supraclinoid stenosis with moyamoya vascular pattern as well as a left parasagittal aneurysm. A subsequent arteriogram revealed a decrease in aneurysm size. Liver function test results were elevated, thyroid-stimulating hormone level was low, and rheumatoid factor (RF),

antinuclear antibody (ANA), erythrocyte sedimentation rate, human chorionic gonadotropin, and leptospira titer were all normal. The acute hospital stay was 26 days and was complicated by fever that was treated as a "CNS infection." She also underwent a shunt replacement for ventriculomegaly.

When she was admitted to rehabilitation, her pupils were round and equally reactive to light, and extraocular muscle movements were intact. A right homonymous hemianopsia and right facial droop were noted. Sensation was normal and motor function appeared to be normal on the left, but assessment was complicated by apraxia. Strength on the right was 3/5, approximately, with increased tone. Hyperreflexia was present on the right. Coordination testing on the left revealed dysmetria. Emotional lability, global aphasia, and apraxia were also present. The FIM score on rehabilitation admission documented impaired locomotion (scale score = 2) and self care skills (scale score = 28). By discharge 23 days later, she was compensating consistently for visual deficits and was using her right upper extremity as a moderate functional assist 50% of the time. Comprehension was improved as well as automatic writing. The patient improved in all areas of functioning, but especially in locomotion (scale score = 12, walking), and self care (scale score = 38).

Case 3

The patient was a 28-year-old right-handed woman with a history of hypertension, hypercholesterolemia, intravenous drug abuse, and a cerebral vascular accident 8 years before this admission with residual left hemiparesis. She had another stroke two years before admission, resulting in right upper extremity paresis and intermittent bilateral upper extremity paresis. An arteriogram revealed severe bilateral supraclinoid ICA and cerebral artery stenosis with a moyamoya pattern. At that time, she presented with complaints of "blindness." Visual evoked potentials were abnormal and consistent with prechiasmatic and postchiasmatic lesions. Magnetic resonance imaging (MRI) confirmed bilateral medial occipital lobe infarcts. Electrolytes (Chem 7), white blood cell, hemoglobin, platelet, partial thromboplastin time, Ca^{++} , Mg^{++} , and phosphate levels were within normal limits. Prothrombin time was mildly elevated (17.6), possibly secondary to nutritional or coagulable dysfunction, and was corrected with vitamin K. Prednisone taper was completed, and carbamazepine, desipramine, and nifedipine were continued.



Fig 1. Angiogram of patient in case 1.

When admitted to rehabilitation, she was blind, had no movement of the eyes toward the left, her pupils were equal, round, and reactive to light, and she had a left facial droop. Gag reflex was intact, but she had limited participation during the examination. Light touch appeared intact except in the left upper extremity. Motor strength was 5/5 in both lower extremities, the right upper extremity, and 3/5 in the left upper extremity. Muscle stretch reflexes were 3+ and symmetric and Babinski's responses were plantigrade bilaterally. She was oriented to person, had perseveration, anomia, and difficulty following single step commands using her right side without physical cues. Right gaze preference with left neglect was present. Frontal release signs were present. Before this admission, she had functional limitations in some areas (table 1), experienced further loss of function, and experienced further prominent deficits in all areas of functioning (self-care = 12, sphincter control = 14, mobility = 6, locomotion = 2, communication = 5, and social-cognition = 5). By discharge 31 days later, she was oriented to person, tone had decreased, and she was compensating for apraxia inconsistently. Motor strength and endurance had improved. The patient regained sphincter control during her admission, but in other areas of functioning, she evidenced limited gains, and at 12-month follow-up, there were apparently no significant changes in FIM scores (table 1).

Case 4

The patient is a 36-year-old woman with hypertension, hypoalbuminemia, congenital single kidney and a ruptured ovarian cyst 15 years before admission, who presented with a history of "depression" of several months. She also experienced acute onset of dysarthria and right hemiparesis. MRI showed bilateral hemorrhagic infarcts of the basal ganglia with a more recent MCA infarction. Antinuclear antibody, rheumatoid factor, thyroid function tests, rapid plasma reagin, human immunodeficiency virus, and Lyme titer results were all normal. Angiogram revealed bilateral supraclinoid narrowing of the ICA. The patient underwent left internal carotid artery angioplasty with improvement in her speech and gait. Two weeks later, her symptoms worsened and a left encephaloduroarteriosynangiosis was performed to restore blood flow via the external carotid artery system. She was treated with a prednisone taper and placed on phenytoin.

When admitted to rehabilitation, her pupils were equal, round, and equally reactive to light. Extraocular movements were intact. With sensory testing, no gross abnormalities were noted and motor strength was 4-5/5 and symmetric. Muscle stretch reflexes were increased in both lower extremities, but there was no clonus or abnormal Babinski's responses. No frontal release signs were present. She was oriented to person and place and could follow three-step commands. Recall of three objects at 5 minutes was normal. FIM scores at the time of admission to rehabilitation revealed mild deficits in sphincter control (scale score = 12), and severe abnormalities in other areas (self-care = 18, mobility = 9, locomotion = 2, communication = 2, and social cognition = 8). By discharge, memory, poor insight, and word finding deficits improved. She was oriented to time, place, and person. The patient made moderate gains during her 53 days in rehabilitation in all areas except sphincter control. At 5-month follow-up, she had achieved minimal additional improvement (table 1).

DISCUSSION

Despite variability in clinical presentation and nomenclature, moyamoya is most frequently evident in the first and third to fourth decade of life. There is a female predominance and adults

usually present with a loss of consciousness, headache, and/or hemiparesis.^{4,5,11,12} CT findings typically document ischemic infarction in children and hemorrhage in adults.^{3,4} Histopathological findings may include concentric intimal hyperplasia in cerebral vessels, fragmentation and lamination of internal elastic lamina, medial and adventitia thinning, lipid deposition in the intima, and internal elastic lamina folding. Although inflammatory changes are typically absent, it is not clear if the primary pathological processes are related to cerebrovascular stenosis or impaired cerebral blood flow secondary to collateral moyamoya vessels.

Treatment strategies have principally involved medical intervention including vasodilators, corticosteroids, and/or agents to reduce blood viscosity or red blood cell agglutination.^{13,14} In contrast, surgical approaches seek to improve blood flow in existing circulation (ie, angioplasty) or through bypassing of the stenosis by connecting external and internal cerebral circulation, most commonly by encephalomyosynangiosis or encephaloduroarteriosynangiosis.³ The former procedure entails peeling the superficial temporal muscle from the temporal bone and placing it on the cerebral surface. In the second procedure, the superficial temporal artery is sutured to the dura mater. Both procedures are intended to increase the blood flow of the external carotid artery to the internal arterial system.

Three of our patients had hemiparesis (two left, one right) as well as dysarthria (two patients) and/or headache or "depression" (one patient each). The fourth patient presented with loss of consciousness and atypical migraines. In the "typical" stroke involving the carotid circulation, hemiparesis (65%) is the most common symptom, headache (20%) the sixth most common, and dysarthria (15%) the eighth most observed symptom.¹⁵ Presentation was similar to "typical" stroke in our small group, but a larger number of patients would help verify this finding.

Of our four patients, two presented with hemorrhagic findings (basal ganglia and choroidal) and two with infarction (left middle cerebral artery, bilateral occipital). The typical stroke pathogenesis includes thrombotic (40%), embolic (30%), lacunar (20%), and hemorrhagic (10%).¹⁵ While thrombotic and embolic strokes usually have ischemic patterns, our limited group had a higher percentage of hemorrhages than expected. Hemorrhage in these patients may be due to the vascular abnormalities including tortuous engorged vessels with softening and necrosis of the media and adventitia layers of the arteries.³

After a "typical" stroke, medical management may include anticoagulation (heparin or warfarin), antiplatelet agents (aspirin, ticlopidine),¹⁵ and, more recently, thrombolytic agents like tissue plasminogen activator.¹⁶ Surgical procedures utilizing carotid endarterectomy have been shown to be effective in appropriate patients.^{17,18} In contrast, there are no guidelines for medical or surgical treatment of moyamoya. The acute treatments for moyamoya involve medical management, surgery to improve the existing circulation, or connecting the external and internal cerebral circulations. Most of the medical treatments for moyamoya have been borrowed from experiences treating "typical" strokes. The surgical approaches are not done in the "typical" stroke because an acute obstruction in the distal artery is commonly responsible for the symptoms, in contrast to moyamoya, where slowly progressive stenosis in a large artery is more amenable to success when an effective resupply of blood is established.

Success of each of these treatments is unknown because long-term outcome data are not readily available. Despite lack of outcome data, medical and/or surgical treatment for moyamoya is available but physicians may not be familiar with treatment options principally because few surgeons have experience with these procedures, eg, encephalomyosynangiosis.

To better understand the functional skills and recovery in moyamoya, FIM scores were completed at admission, discharge from the rehabilitation unit, and during follow-up in the outpatient clinic. A FIM score estimate for prehospital function was determined by history and records, and three patients had the maximum score (or within one point) while one patient (case 3) had prior deficits secondary to moyamoya that were diagnosed a year before the present study. On admission, FIM scores had decreased an average 58 points (43-75) to a mean of 62. Loss of function occurred in all six skill areas except communication and social cognition in case 1, and sphincter control in case 2. Length of inpatient rehabilitation stay was 23 to 53 days (average 34). FIM scores measured during the week of discharge showed gains of 24 to 35 points (average 31). After discharge, FIM scores increased minimally (0 to 7 points) over the 5- to 12-month follow-up. All four patients were discharged home, one with the need for constant supervision secondary to blindness (case 3).

The Uniform Data System for Medical Rehabilitation (UDSMR) for all "first stroke" patients in rehabilitation in 1994 noted that 48% were men with a mean age of 70 years and a mean admission FIM of 62.5. Discharge FIM was 86.6, and length of stay averaged 25 days. Most patients were discharged to the community (76%), but 17% were admitted to long-term care facilities and 5% to acute care. Length of stay efficiency (mean change in FIM score divided by mean days) was .97.¹⁹ In contrast, none of our moyamoya patients were men and the average age (31) was almost 40 years younger than those with stroke. Consequently, these patients will have a disability for a considerably greater number of years than the model stroke patient. Equipment and long-term medical costs will inevitably be higher. Average admission FIM scores were similar in the two groups but discharge FIM scores were higher in the moyamoya group. Higher scores may be due to their longer length of stay, and actually the rate of progress was slower in the moyamoya group with the length of stay efficiency of .88. Also, our patients gained only an additional four points on average during follow-up (5 to 12 months), which suggests they had been discharged after major functional gains had been achieved. Possibly because of the excellent social support available (intact families) and higher functional level at discharge, all of our patients were discharged home. We do not yet have data on long-term survival or functional skill level, but we will follow these cases to gather additional functional and survival data.

Because moyamoya disease is somewhat uncommon, assembling a large clinical sample is difficult. Also, variability in clinical presentation and outcome is apparent even in our small sample. Our patients experienced dramatic impairment in functional skills and considerable but incomplete restitution of function. Moyamoya disease will most likely be associated with residual functional impairment, but these patients may benefit

from rehabilitation oriented towards neurologic deficits, especially if they evidence specific deficits in functional skills (locomotion, self-care, mobility) that may be addressed by therapy and educational components of rehabilitation programs and therapy.

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