



Role of Physiotherapy in Adolescent Moyamoya Disease

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ABSTRACT:

Moyamoya disease (MMD) is a rare, progressive cerebrovascular condition characterized by stenosis of the internal carotid arteries, leading to ischemic and hemorrhagic strokes. Patients often present with motor deficits, balance impairments and reduced functional mobility, necessitating comprehensive rehabilitation to regain independence. This case report involved a 19-year-old female patient with bilateral MMD who underwent encephalo-duroarterio-myo-synangiosis (EDAMS) surgery followed by a two-month postoperative physiotherapy program. Interventions included strength training and balance exercises. Progress was systematically tracked using the Rivermead mobility index and Berg balance scale, with weekly mobility and functional independence assessments. The patient demonstrated significant improvements in mobility and balance. RMI scores improved from 1/15 at baseline to 13/15, while BBS scores increased from 0 to 52 over eight weeks. Structured physiotherapy interventions focusing on balance and strength training have led to marked improvements in postural stability, allowing the patient to regain independent ambulation and perform activities of daily living without assistance. This case report highlighted the critical role of physiotherapy in rehabilitating patients with MMD. Early, individualized interventions can significantly enhance motor recovery, improve balance, and restore functional independence. Continued research is necessary to further optimize rehabilitation protocols for MMD patients.

1. Background

Moyamoya disease (MMD) is a cerebrovascular illness that causes progressive vascular stenosis, blockage of the internal carotid artery (ICA) and the production of excessive, hazy, proximal collateral arteries called "Moyamoya vessels"-(1). This steno-occlusive pattern is connected with the compensatory growth of a collateral network of arteries near the base of the brain, which appears as a "puff of smoke" on conventional angiography ("moyamoya" in Japanese (2)). MMD often affects the intracranial section of the internal carotid arteries, the anterior cerebral arteries, and the proximal region of the middle cerebral artery, while sparing the posterior arteries of the Circle of Willis. (3) In rare situations, this procedure also affects the posterior circulation, which includes the basilar and posterior cerebral arteries (4).

MMD is a rare cerebrovascular illness, with an incidence of around 0.086 per 100,000 people in the US (3).

Epidemiologic studies in Japan estimate the annual incidence of Moyamoya sickness to be 0.35 to 0.94 per 100,000 people (5). According to the current research, the incidence and prevalence rate of this disease and syndrome in India remain unknown. MMD has a peak age of appearance; two-thirds of cases occur in children, while one-quarter of cases are seen in people over the age of 40. Moyamoya disease affects nearly twice as many women as men (6). In general, ischemic stroke affects children, whereas hemorrhagic stroke affects adults (7).

There are suggestions for both congenital and acquired causes, but the exact cause is unknown(8). The clinical presentation and course are determined by the severity and location of vascular stenosis and occlusions, as well as the functionality of the collateral circulation (9). MMD is characterized by ischemia and/or hemorrhagic stroke and cognitive impairment, affecting both children and adults (10). Headaches, involuntary choreiform movements, and motor abnormalities are the most common modes of presentation, affecting 80.5% of the



population. Convulsions occur in approximately 9% of patients (5). Four types of surgical procedures have been described: indirect procedures such as encephaloduroarteriomyosynangiosis (EDAMS), direct revascularization via the superficial temporal artery and the middle cerebral artery (STA-MCA bypass), combined approaches, and, in rare cases, cerebral vasculature denervation. The prognosis of patients with Moyamoya disease is related to age and kind of presentation (11).

Multidisciplinary programs and extensive rehabilitation were conducted. Long-term rehabilitation may help to regain multiple clinical functions. A multidisciplinary rehabilitation team can construct a more appropriate model of motor and cognitive multisensory stimulation to improve consciousness recovery by inducing neural plasticity (12). Physiotherapy interventions including strength training, balance exercises and gait training, are designed to improve motor control, balance and functional mobility in patients who have undergone revascularization surgery. One of the key challenges in rehabilitating patients with Moyamoya disease is improving balance and mobility, as these patients often experience limb weakness, impaired coordination, and reduced functional independence. Balance training is an essential component of physiotherapy that focuses on enhancing stability, coordination, and proprioception, thereby reducing the risk of falls and promoting safe ambulation. It has been shown that structured balance training, along with strength and mobility exercises, can significantly improve postural stability and functional outcomes in patients with neurological impairments (13,14,15).

In this case report, we present an 18-year-old female diagnosed with Moyamoya disease who underwent EDAMS surgery and was referred to physiotherapy for further management due to weakness in her limbs and slurred speech. The role of postoperative rehabilitation, particularly physiotherapy, is crucial to address the resultant motor deficits, enhance functional recovery and improve the quality of life. The intervention strategy began with a thorough evaluation of the patient's baseline functional state using the Rivermead Mobility Scale (RMS), which offered a comprehensive estimate of their present mobility abilities. Therapeutic exercises were tailored to address specific deficits identified during the assessment. These included strength training to

counteract muscle weakness, balance exercises to improve postural stability, Throughout the therapy sessions, close monitoring and periodic reassessment ensured that interventions were effective and adjustments could be made as necessary (14). This case report aims to highlight the effectiveness of a structured physiotherapy program in improving motor outcomes in a young patient with Moyamoya disease.

2. Need of the Study

Most of the study highlights pediatrics and adult moyamoya. Still, very few studies have concentrated on adolescence moyamoya and the results have not been well established. Hence, there is a need to study about adolescent moyamoya and the role of physiotherapy in its management.

3. Case Report

The patient was recruited on 22/05/24 with a known case of Moyamoya disease which was diagnosed previous year where she presented with a sudden onset of numbness in her left thumb in March 2023. Numbness lasted for a few minutes and then spontaneously resolved. However, symptoms recurred intermittently, prompting her to inform her mother. She was taken to a nearby clinic where her grip strength was assessed, showing no signs of muscle weakness. She was prescribed vitamin supplements, but numbness persisted intermittently.

Three days later, she began experiencing difficulty moving her left limbs, which led to an urgent hospital visit. An MRI was performed, revealing acute infarcts in the prefrontal gyrus and deep white matter of the right frontal and parietal lobes. The MRI also showed complete occlusion of the internal carotid arteries on both sides beyond the supraclinoid segments, a finding consistent with Moyamoya disease. Additionally, small diffuse collaterals were noted in the region of the middle cerebral artery (MCA) on both sides, indicating the development of compensatory blood vessels.

Given the diagnosis of Moyamoya disease with a transient ischemic attack (TIA), she underwent encephaloduroarteriosynangiosis (EDAMS) surgery on the right side on May 4, 2023. Following surgery, she received continuous physiotherapy, which helped her recover and resume normal daily activities. She was



advised to return for a similar surgical procedure on the left side after two years.

However, in May 2024, she began to experience new symptoms, including deviation of mouth to left side, mild weakness on her right upper and lower limbs, drooling of saliva and slurred speech. This prompted a second EDAMS surgery on the left side which was done on 19/05/2024. Unfortunately, she suffered a stroke following surgery and was subsequently referred to physiotherapy for comprehensive rehabilitation

4. Procedure

Initial Phase of Therapy: (Week 1)

On first day of physiotherapy, we focused on general exercises aimed at improving the patient's bed mobility. Within two days, patient was shifted to the ward, where noticeable improvements in her mobility were observed. At this stage, our intervention focused on enhancing her balance. Once the patient began to regain her sitting balance through reaching activities and weight-bearing exercises, we progressed to sit-to-stand and standing balance training. Initially, the patient experienced difficulty maintaining balance during these exercises, often losing her stability. However, with continuous therapy, her balance improved, allowing us to advance her rehabilitation.

Progression to Dynamic Standing Balance Training: (Week 2 – Week 4)

After achieving a stable sitting balance, we shifted our focus to dynamic standing balance. The patient was introduced to a series of progressive exercises, including stepping activities, single-leg standing, ball-kicking drills, and strengthening exercises targeting the right side. These activities were designed to improve her confidence and prevent stumbling during ambulation.

Ambulation and Functional Training: (week4-week6)

The patient began walking with two-person support. Remarkably, her facial deviation, a notable neurological deficit, showed significant improvement and normalized with consistent weekly assessments. We introduced more complex gait training exercises, including forward walking, backward walking, sideways walking, and tandem walking. These exercises were performed for 60 minutes, 5 to 6 days a week, with weekly assessments

conducted to evaluate progress and tailor the exercise regimen according to her needs.

Discharge and Continued Rehabilitation: (Week 6-Week 8)

Upon discharge, a comprehensive home exercise program was taught to the patient to continue her rehabilitation journey. Over time, the patient gradually regained comprehension, becoming fully oriented to place, time, and person. Continuous assessments were conducted to monitor her progress, and, eventually, the patient achieved the ability to walk independently without support. She also progressed to climbing stairs and participated in strengthening exercises for her upper limbs during consecutive sessions.

Every week the patient was assessed at the initial day of treatment and following outcome were used to assess patient status. And post-test was done on weekly basis till the end of eight week.

The following outcome measure were used: reflex with a reflex hammer by giving a tap on the tendons, tone by modified Ashworth scale, voluntary control by brunstorm grading, balance with Berg balance scale and mobility comprehensibly assessed with rivermead mobility scale.

5. Results

Table 1: Assessment of Balance and Mobility

| Scales | 22/5/24 | 29/5/24 | 5/6/24 | 12/6/24 | 19/6/24 | 26/6/24 | 3/7/24 | 10/7/24 |
|--------------------------|---------|---------|--------|---------|---------|---------|--------|---------|
| Berg balance scale | 0 | 2 | 8 | 14 | 25 | 32 | 42 | 52 |
| Rivermead Mobility Index | 1/15 | 2/15 | 3/15 | 3/15 | 4/15 | 6/15 | 9/15 | 13/15 |

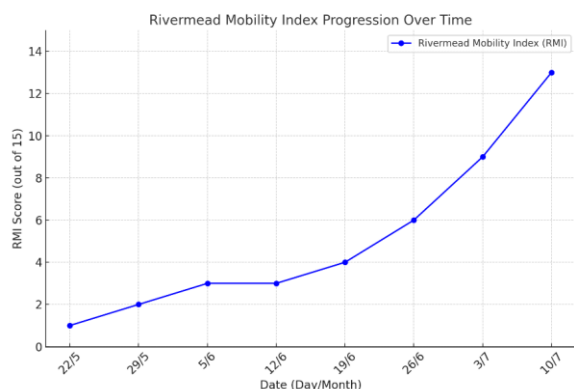


Figure 1: Improvement of mobility over various intervals.

6. Discussion

In this case study, a structured physiotherapy program was implemented for an 18-year-old female with Moyamoya disease (MMD) who underwent bilateral EDAMS surgeries. The focus was on improving motor control, balance, and functional mobility the key areas that often get impaired in patients with MMD. It appears that patients with Moyamoya disease may benefit from rehabilitation oriented toward neurological deficits (15).

This study stands out due to its adolescent subject and inclusion of multiple outcome measures for balance and mobility which were assessed weekly. The RMI, which ranges from 0 to 15 (with higher scores indicating better mobility), showed the patient's initial score at 1/15 on 22/5, reflecting very limited mobility. Over time, consistent improvements were observed, with notable progress after the 5th week (19/6), where the score rose from 3/15 to 4/15. By 3/7, the patient achieved a score of 9/15, indicating significant advancement in various mobility tasks, such as standing unsupported and walking indoors with assistance. By the final assessment on 10/7, the patient reached a score of 13/15, demonstrating substantial improvement in mobility, including the ability to manage uneven surfaces and pick up objects from the floor independently. The Berg Balance Scale (BBS) was utilized to evaluate balance and functional mobility over an 8-week span, revealing a marked improvement in the patient's balance capabilities. The total BBS score increased from 0 at the initial evaluation (22/5) to 52 at the final session (10/7), reflecting considerable progress in both static and dynamic balance tasks. The BBS provided a

comprehensive assessment of mobility improvements over time. Following targeted balance training and mobility exercises, the patient showed significant gains in mobility and functional independence, progressing from complete dependence to a partially independent level by the time of discharge. This progression underscores the effectiveness of the rehabilitation program administered during this period.

This aligns with findings from previous studies, which have shown that early initiation and structured physiotherapy post-revascularization can lead to significant improvements in motor function and overall mobility outcomes (13,14,15).

This case emphasizes the need for continued physiotherapy in the home setting to consolidate the gains achieved during inpatient rehabilitation and further promote functional recovery in adolescents with Moyamoya disease.

Through structured, progressive, and patient-specific physiotherapy interventions, the patient demonstrated significant improvements in balance, mobility, and functional independence. This approach allowed her transition from requiring full support to independent ambulation, highlighting the effectiveness of targeted rehabilitation programs for patients with complex neurological conditions.

7. Conclusion

Physiotherapy is pivotal in managing adolescents with Moyamoya disease, addressing physical impairments, enhancing functional independence and improving the overall quality of life. Through early intervention, individualized rehabilitation programs and a multidisciplinary approach, physiotherapists can significantly impact the recovery and long-term outcomes of these patients. Continued research and innovation in physiotherapy techniques are essential to optimize care and improve the lives of adolescents with Moyamoya disease.

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