

Effect of strengthening exercise and hand functional mobility to reduce muscle atrophy in Hirayama patient.

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ABSTRACT

BACKGROUND Hirayama disease (HD), also known as monomelic amyotrophy or juvenile muscular atrophy of the distal upper extremities, is a unique neurological condition that was initially described by Keizo Hirayama, a Japanese neurologist.

CASE PRESENTATION A 22-year-old man had been experiencing progressive weakness and muscular atrophy in his left hand and forearm for the past six months. He was also having trouble with fine motor skills like gripping, holding items, and finger extension.

INTERVENTION The patient completed a 4 week physiotherapy programme including strengthening and hand functional exercises to improve muscle strength, muscle atrophy and enhance upper limb function.

RESULTS Strength increases (2 to 5 grade) and hand functional mobility improves (5-8). The patient reported no increase in muscle atrophy, better finger extension movement, grasp and release during functional task.

CONCLUSION According to this case study, a patient with Hirayama disease may benefit from an organized physiotherapy program that includes progressive strengthening exercises and hand functional mobility training to enhance upper-limb muscular strength, hand function, and everyday living performance.

KEYWORDS Hirayama disease; Strengthening exercise; Hand functional mobility; Muscle atrophy; Grip strength; Physiotherapy.

BACKGROUND Hirayama disease (HD), also known as monomelic amyotrophy or juvenile muscular atrophy of the distal upper extremities, is a unique neurological condition that was initially described by Keizo Hirayama, a Japanese neurologist.

The juvenile onset of unilateral or asymmetric weakness and amyotrophy of the hand and forearm muscles supplied by myotomes C7–T1, with sparing of the brachioradialis muscles and no objective sensory disturbance or lower-limb involvement, is the hallmark of HD, a regional disorder that is more common in men.

Hirayama disease, also known as monomelic amyotrophy, is an uncommon motor neuron illness that usually starts in adolescence and is characterized by increasing weakening and atrophy in the distal upper extremities.

EPIDEMIOLOGY Hirayama disease is uncommon in clinical settings and has a low incidence. It is uncommon in the Middle East or Arab nations, and the majority of case reports in the literature are from Asian nations. It is more common in Asia and is obviously more common in men, especially in their third decade of life. Just 279 cases have been documented in India over a 35-year span.

ANATOMY Formation: The ventral rami of the upper four cervical nerves combine to form the cervical plexus. The costotransverse bars are grooved by the rami, which appear between the anterior and posterior tubercles of the cervical transverse processes. Three loops are formed by the connections between the four roots. Branches: The lesser occipital nerve (C2), great auricular nerve (C2–C3), transverse cervical nerve (C2–C3), and supraclavicular nerves (C3–C4) are among the superficial branches.

PATHOPHYSIOLOGY

Unbalanced growth in the spinal cord and vertebral column (during adolescent growth spurt)



Tight dural sac + decreased flexibility due to loss of posterior dural attachment



Neck flexion



Posterior dural wall displacement anteriorly



Lower cervical spinal cord compression (C7–C8)



Impaired venous drainage combined with venous congestion (posterior epidural venous plexus)



Prolonged anterior spinal artery microcirculatory insufficiency



Anterior horn cell ischemia that occurs repeatedly ("Poisson effect")



α -motor neuron loss and necrosis



Localized atrophy and gliosis of the lower cervical spinal cord



Hand and forearm muscle weakness and atrophy (Hirayama illness)

AUTOPSY FINDINGS:

Chronic ischemia



>50% decrease in cervical cord AP diameter



Central necrosis



Absence of macrophage infiltration and loss of both large and small anterior horn cells



Atrophy of the lower cervical cord (C7-C8)



Weakness and atrophy of the hand and forearm muscles

ETIOLOGY The pathophysiology of this phenomenon has been the subject of numerous theories. Degenerative spurs during flexion compress the cord, according to the contact pressure theory, which was deduced from cadaveric research in 1960. In 1987, the theory of the tight dural canal in flexion was put out. This theory, which is the most widely acknowledged, predicts forward cord movement during cervical flexion by describing a greater laxity of the dura mater from its superior anchors on the dorsal surfaces of C2 and C3. During cervical flexion, increased laxity causes the spinal cord to move forward, resulting in microischemia in the anterior horn and recurrent subclinical damage. Asymmetric lower cervical cord thinning within the C7 and C8 shows that trauma and microischemia ultimately result in myelopathy and degeneration.

CLINICAL FEATURES The compilation of the prevalent clinical manifestations of HD, which include five main symptoms and manifestations, has been made possible in the last ten years in particular by the ongoing improvement in clinicians' awareness of the disease and the numerous pertinent studies: **1.** Distal weakness and wasting in one upper extremity or asymmetrically in both upper extremities, primarily on the ulnar side. The majority of patients exhibit more severe ulnar muscle atrophy, a condition known as "reverse split hand syndrome," which may be a characteristic that sets HD apart from ALS. This kind of injury is most likely to affect the AHCS that innervate the hypothenar muscles.**2.** The disease has an

insidious onset and progresses for three to five years before either stopping or going thru a comparatively benign phase.³ The affected hand's fingers tremble irregularly and coarsely.⁴ When a person is in a cold environment, their symptoms slightly aggravate for a brief period of time.

INVESTIGATION The most crucial imaging test for the diagnosis of Hirayama disease (HD) is neck-flexion MRI. The aberrant tightness of the dural sac during cervical flexion indicates the pathophysiology of HD and compression of the lower cervical spinal cord. A crescent-shaped high-intensity mass, cord flattening, atrophy, and forward displacement of the posterior wall of the cervical dural sac are among the symptoms. Electrophysiological tests are crucial for HD diagnosis and distinction. Without problems with the sensory nerves, the primary symptoms of EMG include segmental neurogenic injury of the AHCs or anterior root of the spinal nerves in the lower cervical spinal cord.

TREATMENT Non-surgical: For many patients, cervical collars are the first-line non-surgical treatment for stopping the disease's progression. By limiting neck movement and preventing compression injury to the spinal cord during flexion, this approach stops HD from getting worse. Surgical: In addition to relieving subjective symptoms, surgery also improves objective examination results. On the one hand, clinical signs and EMG, the absence of venous congestion recurrence, and improved grip strength due to the recovery of muscles other than the atrophic intrinsic muscles demonstrate that surgery reduces the natural duration of the illness.

AIM: Effect of strengthening exercise and hand functional mobility to reduce muscle atrophy in Hirayama patient.

OBJECTIVE:-

- To evaluate the severity of muscle atrophy in the affected upper limb.
- To evaluate hand movement accuracy and coordination
- To evaluate functional mobility of the hands both before and after the intervention.
- To evaluate the affected upper limb's muscle strength.
- To evaluate the strength of the hand grip.

CASE DESCRIPTION A 22-year-old man had been experiencing significant weakness and muscular atrophy in his left hand and forearm for the past six months. He was also having trouble with fine motor skills including gripping, holding objects, and extending his fingers. The patient complained of weakened hand grips, tired muscles, and trouble using their hands functionally. Using the afflicted hand for extended periods of time made the symptoms worse. No noticeable discomfort or loss of sensation was observed.

clinical examination, the intrinsic hand muscles, FDI (first dorsal interosseous), and forearm muscles were found to be visibly atrophied. Additionally, there was a decrease in hand functional mobility, muscle strength, and fine motor coordination. A cervical spine MRI revealed modest lower cervical cord atrophy (C5–C6) and anterior displacement of the posterior dural sac with neck flexion. A localized neurogenic pattern involving the left upper limb muscles was revealed by EMG and nerve conduction investigations, suggesting long-term denervation and reinnervation alterations. Research on nerve conduction was normal.

UNIQUENESS OF THE STUDY Rare neurological condition: There aren't many physiotherapy case studies that concentrate on the rehabilitation of Hirayama disease, an unusual cervical myelopathy.

Conservative physiotherapy approach: The study highlights the use of hand functional mobility training and strengthening exercises for non-surgical care, which is less commonly reported than medical or surgical treatment.

Focus on reducing muscle atrophy: Thru a planned exercise regimen, the intervention seeks to improve muscle strength and slow the progression of muscle wasting rather than only preserving function.

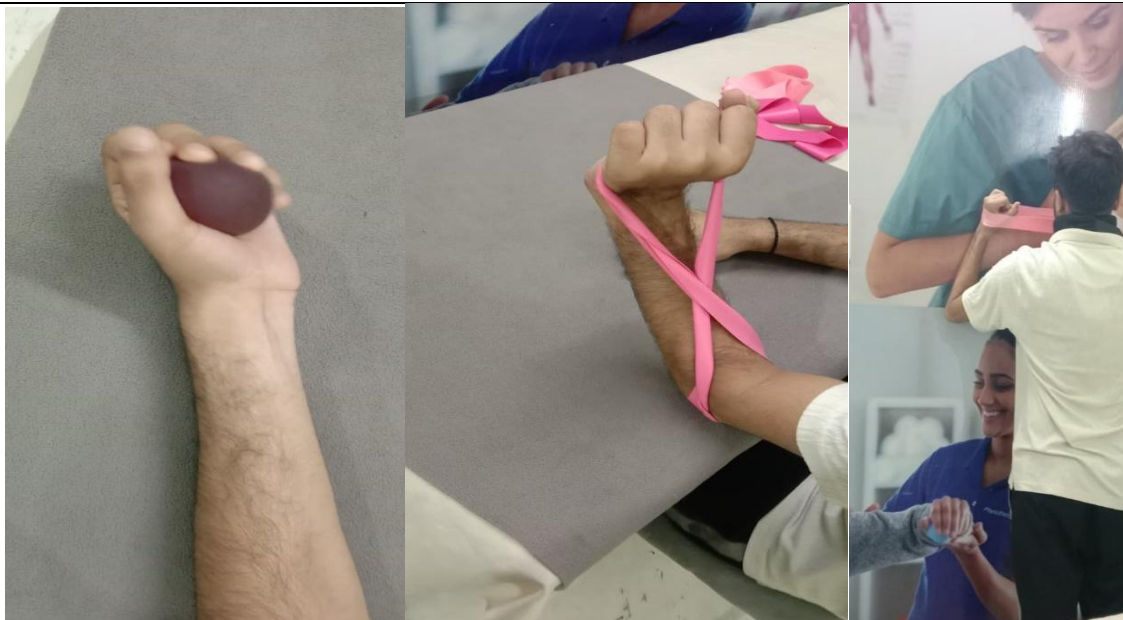
Improvement in hand function: The most often impacted functions in Hirayama disease are hand grip strength, dexterity, coordination, and functional activities, all of which are specifically evaluated in this study.

INTERVENTION 1.Strengthening exercises: Strengthening activities were used to increase upper limb function, decrease muscle atrophy, and increase muscle strength. The program included low-resistance progressive exercises to target the hand, wrist, forearm, elbow, and shoulder muscles. Patient position: The patient was comfortably seated in a chair with support for their back. The afflicted upper limb was positioned

comfortably on a treatment table. Therapist position: Standing next to the afflicted upper limb, the physiotherapist guided the movement, stabilized the limb, and offered resistance or physical support as needed. Method: Progressive Resistance Exercise (PRE). Technique: A theraband, a soft ball, and light dumbbells (0.5–1 kg) were used to perform active resisted workouts after isometric contraction of the wrist extensors, finger extensors, biceps, triceps, and shoulder muscles. Each exercise was done in two to three sets of ten to fifteen repetitions, with enough rest in between. The patient's tolerance was taken into consideration when progressively increasing resistance. **2. Hand functional mobility training**: To enhance hand dexterity, grip strength, fine motor coordination, and independence in daily living tasks, hand functional mobility training was offered. Patient position: The patient was sat comfortably, with the hand positioned for functional activities and the forearm resting on a table. Therapist position: The physiotherapist supervised the patient's performance, stood or sat next to them, and gave verbal and manual instructions as needed. Method: Task-oriented functional training. Technique: The patient engaged in object grasp-and-release training, pebble picking, coin picking, finger abduction/adduction, and grip strengthening with a soft ball and finger opposition exercises. With breaks as needed, each exercise was repeated for ten to fifteen minutes.

RESULT After receiving physical treatment for four weeks, the patient's upper limb function improved. During functional tasks, the patient's hand grip strength improved and they showed improved finger extension movement, grasp, and release. Improved hand functional mobility made it simpler to do tasks like holding a cup and picking up tiny objects. Additionally, coordination and endurance improved, and weariness during repetitive tasks decreased. During the course of treatment, neither an increase in muscular atrophy nor a worsening of neurological symptoms was noted.

Outcomes measure	Pre-treatment	Post-treatment
MMT SCALE	Grade 2/3	Grade 5
(PSFS) Patient-Specific functional scale		
Holding a cup	5/10	8/10



DISCUSSION In 2020, Tinu Sethi, Sushma Rawat, S. L. Munde, This case study showed that hand grip strength, muscle endurance, dexterity, and functional performance were all greatly enhanced by an organized strengthening training program. Exercise decreased functional impairment and improved performance of daily living activities, even tho muscle bulk did not fully recover. In 2022, Hongwei Wang, Ye Tian, Jianwei Wu, Sushan Luo, Chaojun Zheng, Chi Sun, Cong Nie, Xinlei Xia, et al. The review emphasized that the first-line strategy for Hirayama disease is still conservative treatment. Muscle performance and functional capacity are enhanced by physiotherapy that emphasizes hand function training, posture correction, cervical stabilization, and muscle strengthening. Early intervention improves the patient's quality of life and reduces the progression of the condition. In 2024, Sara Lay, Aashrai Gudlavallesi,

Sunil Munakomi, Sandeep Sharma, According to the authors, Hirayama illness is a self-limiting lower motor neuron ailment that causes hand and forearm muscular atrophy and weakening. They underlined that early conservative treatment, such as the use of cervical collars and physical therapy, helps prevent more muscle atrophy, maintain functional independence, and retain muscular strength. To improve everyday tasks and stop advancement, regular exercise and rehabilitation are crucial.

The current study's results are in line with the previously mentioned literature. The patient participated in a rehabilitation program that included hand functional mobility training and strengthening exercises. Hand muscular strength, grip function, finger movements, coordination, and performance of everyday functional activities all showed improvements after the training. Additionally, the patient's ability to control their upper limbs improved and they had less trouble with fine motor activities. These results imply that hand functional mobility training in conjunction with strengthening exercises can minimize functional deterioration, lessen the effects of muscle atrophy, and enhance total hand function in individuals with Hirayama illness.

CONCLUSION This case study indicates that a patient with Hirayama illness may benefit from a systematic physiotherapy program that includes progressive strengthening exercises and hand functional mobility training to enhance upper limb muscular strength, hand function, and performance of daily living tasks. Targeted rehabilitation can assist delay functional deterioration, increase residual muscular performance, and improve the patient's quality of life even tho muscle atrophy brought on by Hirayama illness might not be entirely reversible. To maximize functional independence and avoid secondary problems, early diagnosis, frequent follow-up, and tailored physiotherapy interventions are crucial.

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