

Title: The Role of Ergonomics in Enhancing Rehabilitation Outcomes in Hirayama**Disease: A Case report****Running title:** Ergonomic intervention for Hirayama disease**Authors:** Dr. Nupoor Kulkarni¹, Aashirwad Mahajan², Ankita Yelikar³, Sakshi Dupare⁴, Rupinder Bhatti⁵

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Total pages: 9**Abstract**

Introduction : Hirayama disease (HD) is a rare, self-limiting cervical myelopathy characterized by progressive distal upper limb weakness and muscular atrophy, predominantly affecting young males which can significantly impact occupational and daily activities. This study was done to examine the effectiveness of a physiotherapy and ergonomic interventions in a patient with HD.

Case Description: A 22-year-old male printing professional diagnosed with HD presented with progressive weakness of the left hand and forearm with recent involvement of the right hand, resulting in difficulty performing work-related tasks. Clinical examination revealed bilateral distal upper limb weakness and muscle wasting (left greater than right), and preserved sensation. Dynamic cervical MRI demonstrated cord thinning at C5–C7 with anterior displacement of the posterior dura during neck flexion.

Intervention: The patient underwent a 6-week rehabilitation program consisting of faradic stimulation, pneumatic glove-assisted hand training, cervical and upper limb strengthening exercises, fine motor dexterity training, and individualized ergonomic modifications, including posture correction, workstation redesign, cervical collar use, adaptive writing aids, and education to avoid prolonged neck flexion.

Results: Following intervention, Purdue Pegboard scores improved bilaterally (Left: 9–13; Right: 11–15), grip strength increased (Left: 16.2–21.7 kg; Right: 18.5–24.8 kg), wrist range of motion improved in all planes, and Patient-Specific Functional Scale scores increased from 3.0 to 7.3.

Conclusion: A comprehensive rehabilitation approach having both physiotherapy and ergonomic interventions may improve grip strength, mobility, and functional independence promoting continued occupational involvement in individual with Hirayama disease.

Keywords: cervical myelopathy; Hand dexterity; Grip strength; Occupational performance.

Introduction: Hirayama disease (HD), also known as monomelic amyotrophy or juvenile muscular atrophy of the distal upper extremity, is a rare cervical myelopathy that predominantly affects adolescent and young adult males. First described by Hirayama in 1959, the condition is characterized by insidious onset of unilateral or asymmetrically bilateral weakness and wasting of the distal upper limb muscles, particularly those innervated by the C7–T1 myotomes, while classically sparing the brachioradialis muscle, resulting in the characteristic appearance of “oblique amyotrophy.” The disease typically progresses over a period of 1–5 years before reaching a spontaneous plateau, distinguishing it from progressive motor neuron disorders such as amyotrophic lateral sclerosis (ALS).¹

The current understanding of HD pathophysiology is centered on a dynamic cervical cord compression mechanism. During adolescence, disproportionate growth between the vertebral column and the dural sac may result in a relatively tight cervical dural canal. Consequently, neck flexion produces anterior displacement of the posterior dural wall, leading to repeated compression of the lower cervical spinal cord. This dynamic compression is associated with venous congestion and chronic microcirculatory insufficiency in the territory of the anterior spinal artery, particularly affecting the anterior horn cells at the C7–T1 levels. Repeated ischemic insults are believed to result in selective motor neuron degeneration, thereby explaining the characteristic distal upper limb weakness and atrophy observed in affected individuals.¹

Accurate diagnosis of HD requires a high index of clinical suspicion, particularly in young males presenting with distal upper limb wasting in the absence of sensory deficits or upper motor neuron signs. Electrophysiological studies may reveal characteristic findings, including the reverse split-hand pattern, wherein the compound muscle action potential (CMAP) of the abductor digiti minimi is disproportionately reduced compared with that of the abductor pollicis brevis. This pattern may aid in differentiating HD from ALS and other motor neuron disorders. Furthermore, routine cervical MRI obtained in a neutral position may appear

normal or demonstrate only subtle lower cervical cord atrophy. Dynamic flexion MRI remains the imaging modality of choice, as it can demonstrate anterior displacement of the posterior dural sac, widening of the posterior epidural space, and engorgement of the posterior internal vertebral venous plexus, findings considered highly characteristic of HD.²

Although HD is generally considered a self-limiting disorder with eventual disease stabilization, the resultant impairments in strength, dexterity, and upper extremity function can substantially affect activities of daily living, educational pursuits, occupational performance, and overall quality of life.³

Current evidence regarding physiotherapy management in HD remains limited and is largely confined to case reports and small observational studies. Nevertheless, emerging literature suggests that targeted rehabilitation interventions can positively influence upper limb function and disability outcomes. Consequently, management strategies should extend beyond prevention of neurological progression and focus on optimizing functional capacity, maintaining independence, and minimizing secondary biomechanical complications.

Aim: To examine the effectiveness of physiotherapy and ergonomics in optimizing function, strength, flexibility.

Patient Information:

A 22-year-old male from printing profession, diagnosed with Hirayama disease was referred to physiotherapy department with complaints of slow progressive weakness of left hand, and forearm, with an insidious onset and starting of involvement of right hand from past two months, difficulty at workplace to continue the job.

C/C: Difficulty while holding objects, lifting weights

Diagnostic Assessment and clinical findings:

- MRI showed thinning of cord at C5-C6-C7 level with forward shift of post dura exaggerating on flexion study, Loss of normal cervical lordosis.
- Apparent atrophy in thenar, hypothenar, interosseous muscle and forearm muscles L>R. Occasional numbness and chills in his left arm. Minimyoclonus +, diminished 1+ reflexes in left UL, sensations intact.
- Purdue **Pegboard Test (PPT)**: showed impaired fine motor dexterity, hand–eye coordination, and manual performance of the affected upper extremity.

- **Grip Strength:** Assessed using a handheld dynamometer showed diminished hand muscle strength and functional grasp capacity.
- **Range of Motion (ROM):** Active wrist ROM was measured using a goniometer for: Wrist flexion, extension, Radial deviation, Ulnar deviation which was reduced.
- **Patient-Specific Functional Scale (PSFS):** Used to evaluate patient-perceived difficulty in performing individualized functional activities and to monitor functional improvement over time. Pre-intervention, the patient reported diminished functional status.

Therapeutic Intervention:

Six weeks intervention consisted of:

1. **Electrical Muscle Stimulation (Faradic Current)** Stimulation to wrist extensors, finger extensors, and intrinsic hand muscles for 50 contractions each for 5 days/week for 6 weeks.⁵
2. **Pneumatic Glove Training:** Repetitive assisted finger flexion-extension movements using pneumatic glove device 10 minutes/session 5 days/week for 6 weeks
3. **Exercises:** Cervical ROM Exercises, Shoulder muscles Strengthening Exercises, Elbow muscles Strengthening Exercises, Wrist Hand and Finger Strengthening Exercises 3 sets × 10 repetitions, 3 days/week for 6 weeks^{2,6,7}
4. **Fine Motor Dexterity Training** in the form of Pegboard activities, coin manipulation, buttoning/unbuttoning, writing practice 5-10 minutes/session, 5 days/week for 6 weeks²
5. **Ergonomic Advice & Environmental Modifications**
 - a. **Cervical Spine Care**

Neck position: Avoid prolonged flexion during reading, studying, or using a mobile phone. •Postural re-education and avoidance of prolonged neck flexion (studies, mobile phone use).
 - b. **Desk setup:**

Books/computer screen kept at **eye level** with a stand or height-adjustable table. Use of a **tilted board** to minimize head bending.

Cervical collar: Lightweight cervical collar during work to limit excessive flexion. Cervical collar advised during activities to minimize neck flexion and reduce cord compression.

Mobile usage: Use a mobile stand instead of looking down at the phone.

c. Workstation modification:

Seating: Chair with good lumbar and cervical support, height adjusted so that forearms rest comfortably on the table.

Lighting: Proper illumination to avoid leaning forward.

Voice typing or speech-to-text software for prolonged assignments.

d. Hand Function & Writing Modifications

Adaptive pens: Use **pen grips, triangular pens, or fat markers** to reduce fine motor strain.

Writing boards/tables: Smooth surface with adequate elbow support.

Keyboard modifications: Ergonomic keyboard and mouse to reduce repetitive strain.

- e. **“20–20–20 rule”** – every 20 minutes, look 20 feet away for 20 seconds, and take posture breaks.^{8,9}

Results:

The results stated that:

1. Purdue Pegboard Test: The Purdue Pegboard Test showed improvement in fine motor dexterity and hand coordination after 6 weeks of treatment. (Table 1) This improvement indicates better finger agility, manual dexterity, and hand function required for daily activities such as writing, buttoning clothes, and handling small objects.

2. Grip Strength: The Grip Strength was significantly increased in hand muscles following the intervention. (Table 1) These findings suggest enhanced muscular performance, improved grasping ability, and better functional use of the hands in activities requiring forceful gripping.

3. Wrist ROM: The Range of Motion (ROM) graph illustrates substantial improvements in wrist mobility bilaterally over the 6-week treatment period. (Table 1) These improvements indicate reduced joint stiffness, increased flexibility, and enhanced functional movement of the wrist, contributing to better performance of daily activities.

4. Patient-Specific Functional Scale (PSFS): The PSFS graph shows a marked improvement in the patient's perceived functional ability. (Table 1) This indicates that the

patient experienced a substantial reduction in activity limitations and was able to perform personally important tasks more effectively following treatment.

Table 1: Pre-post scores of outcome measures

S.N	Outcome	Left		Diff	Right		Diff
		Pre	Post		Pre	Post	
1	Purdue Pegboard Test	9	13	4	11	15	4
2	Grip strength	16.2	21.7	5.5	18.5	24.8	6.3
3	ROM						
	Wrist flexion	50	75	25	45	65	20
	Wrist extension	45	70	25	40	65	25
	Radial deviation	12	20	8	10	18	8
	Ulnar deviation	22	38	16	20	35	15
4	PSFS Patient-Specific Functional Scale (PSFS)	3	7.3	4.3	3	7.3	4.3

Discussion:

The present case highlights the importance of early diagnosis and comprehensive rehabilitation in Hirayama disease (HD), a rare cervical myelopathy characterized by progressive distal upper limb weakness and muscular atrophy. Improvements in Patient Specific Functional Scale, Purdue Pegboard Test, Manual Muscle Testing (MMT) Strength grading of wrist flexors/extensors, intrinsic hand muscles, Grip Strength, Range of Motion for wrist and fingers have been reported after completion of intervention.

The role of cervical collar use also warrants emphasis. Consistent with previous reports, cervical orthosis remains one of the few interventions shown to potentially limit disease progression by restricting excessive neck flexion and reducing repetitive anterior displacement of the posterior dural sac. However, the effectiveness of cervical

immobilization may be enhanced when combined with ergonomic education and behavioral modification strategies that encourage adherence to neck-protective postures during work, study, and leisure activities. Thus, patient education regarding posture, workstation ergonomics, activity pacing, and avoidance of sustained cervical flexion should be regarded as a core component of long-term management. This findings support the hypothesis that enhanced cervical stability may reduce biomechanical stress on the cervical spinal cord and improve neuromuscular efficiency.⁴

The multimodal rehabilitation program incorporated physiotherapeutic modalities like faradic stimulation, pneumatic glove, strength and flexibility exercises and individualized ergonomic intervention. Particular emphasis was placed on optimizing sitting posture, workstation configuration, reading and writing habits, smartphone use, and activity modification to minimize prolonged neck flexion. These interventions were designed to promote neutral cervical alignment during functional activities and reduce cumulative biomechanical loading on the lower cervical spine.^{5,6,7}

The rationale for integrating ergonomics with exercise therapy is supported by evidence demonstrating that spinal alignment influences neuromuscular control, sensorimotor integration, muscle activation patterns, and movement efficiency. Poor postural habits and sustained flexed-neck positions may increase mechanical strain on cervical tissues, potentially exacerbating neurological compromise and secondary musculoskeletal dysfunction. Ergonomic interventions can therefore complement therapeutic exercise by reinforcing optimal movement patterns throughout daily activities, thereby extending the benefits of rehabilitation beyond the clinical setting.⁸

A distinctive aspect of the current study was the integration of ergonomic intervention alongside therapeutic exercise. Ergonomics, defined as the scientific discipline concerned with understanding interactions between individuals and their environments to optimize performance, safety, and well-being, is particularly relevant in HD because excessive or sustained cervical flexion has been implicated in disease pathomechanics. Therefore, ergonomic modification represents not merely a supportive intervention but a critical component of comprehensive rehabilitation aimed at reducing repetitive mechanical stress on vulnerable cervical structures. Evidence from occupational health research indicates that ergonomically optimized environments are associated with reduced injury risk, improved work performance, enhanced satisfaction, and greater productivity. In the context of

Hirayama disease, where repetitive and sustained cervical flexion may contribute to neurological compromise, ergonomic modification assumes additional clinical significance. Strategies aimed at optimizing posture, workstation configuration, and activity-specific biomechanics may help minimize cervical loading, reinforce neck-protective behaviors, and support long-term functional independence. Therefore, ergonomics should be viewed not merely as an adjunct to physiotherapy but as an integral component of patient-centered management designed to maximize participation and occupational well-being.⁸

From a rehabilitation perspective, ergonomics serves as a bridge between impairment-based interventions and real-world functional performance. By facilitating adaptation of the physical environment to the patient's capabilities, ergonomic strategies can enhance occupational efficiency, reduce fatigue, improve safety, and promote long-term independence. Such interventions are particularly important in young individuals with HD, for whom preservation of educational and vocational participation remains a major treatment goal.⁹

Several limitations should be acknowledged. As a single-case report, the findings cannot be generalized to the broader HD population. The absence of long-term follow-up precludes conclusions regarding the sustainability of treatment effects, and the individual contributions of specific rehabilitation components, including NMES and ergonomic interventions, cannot be isolated. Furthermore, standardized physiotherapy and ergonomic management protocols for HD have not yet been established, reflecting the broader lack of controlled clinical studies in this field. In future, the development of evidence-based rehabilitation and ergonomic guidelines may facilitate more consistent management of this rare but potentially disabling condition.

Conclusion: From the present case it was concluded that a comprehensive rehabilitation approach that integrated physiotherapy and ergonomics was beneficial in the recovery of Hirayama disease. Early diagnosis followed by patient-centered rehabilitation and ergonomic intervention may therefore play a pivotal role in maximizing long-term functional and occupational outcomes in individuals with HD.

Patient perspective: “Before treatment, I had difficulty using my hands for work and daily activities. After the rehabilitation program and ergonomic modifications, I noticed improved ability to hold objects and perform tasks, and greater confidence in continuing my work”

Informed Consent: A written informed consent was obtained from the patient before the treatment and included permissions related to data usage for scientific publications.

Ethical Clearance: Ethical clearance was taken from Institutional Review Board

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Conflict of interest - Nil

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