

## Effect of Strengthening Exercises and Muscle Stimulator in Hirayama Disease Patients Having Muscle Atrophy and Weakness : A Case Report

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### INTRODUCTION

Hirayama disease is juvenile muscular atrophy of distal upper extremity. It is a rare neurological disease characterized by insidious unilateral or bilateral muscular atrophy and weakness of the forearm and hands without sensory or pyramidal signs. This condition was first described by and named after, Hirayama in 1959.<sup>1,2</sup> These cases are mostly seen in asia.

### CASE REPORT

A patient [X], 25 Y/M from Kathmandu, Nepal complains of difficulty in gripping, atrophy of forearm and hand. Patient weakness started 2 years ago with difficulty in thumb movement progressing to the other finger and forearm with tingling sensation on left hand. Firstly, the symptom of tingling sensation was present in the left little finger and over the year it progresses proximally. After a year muscle atrophy was noticed in the hypothenar area.

### ABSTRACT

Hirayama disease, a rare neurological disease, is characterized by insidious unilateral or bilateral muscular atrophy and weakness of the forearms and hands, without sensory or pyramidal signs. The objective of the study is to check the effect of physiotherapy intervention in this rare disease. Three cases with the weakness in grip and atrophy of forearm and hand was given the treatment using muscle stimulator, pressure biofeedback cuff, theraputty and strengthening exercise using weight and were performed 6 days a week for 8 weeks. Neck flexors strength was measured by using pressure biofeedback and upper limb strength was tested through Manual Muscle Testing grades. The deep neck flexors strength were improved using biofeedback cuff and muscle strength of forearm was improved by the resistive exercise using weight which was measured by MMT grading. Grip strength was also improved using theraputty and grip strengthening exercises. Physiotherapy Interventions can prevents the muscle wasting, joint stiffness and improves the muscle strength which significantly leads to improved quality of life.

**Keywords:** Biofeedback cuff; Muscle atrophy; Muscle stimulator; Strengthening exercise; Weakness.

### ASSESSMENT AND EVALUATION

On examination, the subjects was concious, co operative and well oriented. On sensory examination, all the deep and superficial were intact. On motor examination, tone of all the limbs were normal but there was atrophy noticed on the left upper limb in the thenar, hypothenar, interosseous and ulnar side of forearm. Jamar handheld dynamometer measured the strength of grip on left (8lbs) and on the right (30lbs). Pressure Biofeedback Unit was used to assess the strength of neck flexors (25mmHg \* 5 rep \* 10 sec hold ).

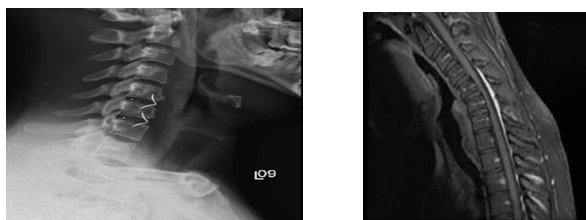


Figure 1: Cervical spine X-ray showing lordosis.

The forearm strength was 3 on MMT scale. All blood investigations were normal. Cervical spine X-ray shows loss of cervical lordosis. Cervical spine X-ray shows loss of cervical lordosis. MRI in flexion position shows there is stretching of the posterior dura, anterior dural shift, and compression of the spinal cord at level C5 and C6.

## INTERVENTIONS

The application of a soft collar was suggested which helps in biomechanical correction required to limit the disease. Postural correction and reeducation were taught to avoid further progression. For strengthening of deep neck flexor muscle towel press exercise was given. Patient was asked to lie supine on the bed and the towel was kept in the lower occipital region and was asked to press the towel with the force of the neck muscles. Patient was instructed to not move the head or to use the hand or body to use force. This exercise was done for 10 repetition with 10 second hold in each repetition. Secondly, isometrics exercises for neck was taught. Patient was asked to put the hand on the forehead, back of head, on the left and right side and to give the pressure on the hand by the head with keeping the spine erect and head neutral. This exercise was done 4 times a day repeating 10 times in each session with hold it 10 second on each doing. Thirdly, Pressure Biofeedback Unit (PBU) was used to strengthen the deep flexor muscles. Patient was asked to lie supine on bed and the unit was placed upper cervical region. The visual unit of PBU was placed in the one hand and the pressure was applied to the PBU with body being neutral. This maneuver is done with 10 repetition holding 10 sec in each doing for 3 times a day. Hand held dynamometer and therapy was used to increase the strength of the forearm and hand. It was done for 4 times a day with 40 repetition each. Muscle stimulation was used on the motor points of forearm of left hand. One point of stimulator was kept on the C7 region and the other one was kept on the motor points of pronator teres, flexor digitorum superficialis, pronator quadratus, extensor digitorum, extensor carpi ulnaris. Each point was given 10 minutes of faradic stimulation for one time a day.

Patient improvement was significant over the period of 8 weeks. Baseline neck flexor muscle strength was 25 mmHg (10rep\*10sec) and over the period of 8 weeks it was increased to 45 mmHg (10 rep\*10 sec). Similarly, strength of fore arm was measure through Manual Muscle Testing (MMT) was 3 on the initial days and was improve to 4+ on later stage. Grip strength was improved to 25 lbs from 8 lbs. The improvement was significant and quality of life of patient was improved amazingly.

## DISCUSSION

HD is a cervical myelopathy affecting juvenile people characterized by localized amyotrophy in the forearm and hand.<sup>3</sup> Its progressive and in our case patient had history of 2 years We used muscle stimulator with combination of strengthening exercises for 8 weeks which improved the atrophy and strength from the early 3 weeks compared to study done in India only with exercises where the improvement was not so significant after 5th week.

Normally, the spinal dura mater is loosely attached to the vertebral canal by the nerve roots, the periosteum at the foramen magnum, the dorsal surfaces of C2 and C3, and the coccyx. The dura mater is loose enough to adjust with the increased length of the cervical spine during the flexion movement of the cervical spine. In patients with HD, the taut dural sac separates the posterior dural sac from its adjacent lamina and on neck flexion, cannot compensate for the increased length of the posterior wall. Thus, the posterior dural wall shifts anteriorly, compressing the cervical spinal cord against the posterior margin of adjacent vertebral bodies.

This compressive force also affects the anterior spinal artery, leading to impaired microcirculatory disturbances in its territory in the lower cervical cord, and the anterior horn cells which are vulnerable to ischemia begin to degenerate, resulting in localized cord atrophy of the lower cervical region, weak and wasted hands, and forearms. Similar cases have been reported worldwide. The findings for this report were similar to the diagnostic criteria of HD according to Tashiro et al.<sup>4</sup> described in the literature.<sup>5,6</sup>

To date, no study has been published regarding a specific treatment protocol for this rare disorder with combination of muscle stimulator and strengthening exercises. This is one of the detailed assessment and evaluation of this specific rare disorder and also establishes general physiotherapeutic interventions. Strengthening exercises and muscle stimulator are helpful in limiting muscle weakness and increasing overall strength, maintaining muscle properties, and improving overall outcomes. The exercise were performed 3-4 times a day for 8weeks [6 days a week] and there is a distinct difference in a patient strength of forearm and grip just after the 3rd week which is a milestone.

## CONCLUSION

Muscle stimulator with strengthening exercise provides early improvement in HD. Early diagnosis of HD with physiotherapy intervention helps in further progression and early improvement with better active daily living.

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