



Case Study

Physical therapy intervention with a low frequency of exercise for a patient with a complicated form of hereditary spastic paraplegia: a case report

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Abstract. [Purpose] Hereditary spastic paraplegia (HSP) is a neurodegenerative disorder characterized by progressive spasticity and weakness of the lower limbs. To date, the appropriate frequency and intensity of physical therapy for patients with HSP are not well-known. We created an original rehabilitation program for a patient with a complicated form of HSP, wherein the program required low-frequency involvement to adapt to the long-term insurance system in Japan. We wanted to find out whether this program could maintain the physical functions and activities of daily living (ADL) of the patient. [Participant and Methods] A 41-year-old male diagnosed with a complicated form of HSP with decreased visual acuity and ataxia of the trunk and upper limb underwent a specific rehabilitation program that included a squatting exercise, a kneeling position exercise, and a motion exercise of taking a bath. This intervention program lasted for 20 minutes per session, with a frequency of two days per week. The patient was in the program for 12 weeks. [Results] All outcome measures, including muscle strength (grip force and quadriceps) and Barthel index, remained unchanged at the end of the intervention program. [Conclusion] The original intervention program used in this study, which had a low frequency of exercise, was effective in preventing further regression of the lower limb function of the patient with a complicated form of HSP, and in preventing a decrease in the ability of the patient to perform ADL.

Key words: Complicated form of hereditary spastic paraplegia, Low exercise frequency, Maintenance of ADL

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INTRODUCTION

Hereditary spastic paraplegia (HSP) consists of a clinically and genetically heterogeneous group of neurodegenerative disorders characterized by progressive spasticity and weakness of the lower extremities¹⁾. According to several epidemiological surveys for HSP in Japan, the prevalence rate of HSP in Japan is estimated to be approximately 0.2 per 100,000 populations²⁾.

Clinical types of HSP are classified into pure and complicated forms³⁾. In the complicated form^{4, 5)}, the degenerative process affects multiple parts of the nervous systems resulting in various neurological symptoms such as cognitive impairment, seizures, extrapyramidal involvement, ataxia, optic atrophy, neuropathy, and muscular atrophy.

Concerning physical therapy for HSP, gait patterns⁶⁾, measurement of functional evaluation⁷⁾, and disease severity⁸⁾ of HSP have been the major interest, and only a few studies were performed regarding physical therapy intervention for HSP. To our knowledge, there has been no published literature that described the frequency, intensity and details of physical therapy for

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the maintenance of the function of lower extremities and Activities of Daily Living (ADL) in HSP patients. Only Asir et al.⁹ proposed an original 8-week intensive rehabilitation program for pure type HSP from which the functional improvement was obtained. Their program consisted of 60–90 min exercise for each session and the frequency of exercise was 6 days per week.

We had an opportunity to provide rehabilitation to a patient with complicated form of HSP. We developed an original physical therapy intervention program with low exercise frequency adapted to the limitations of the long-term care insurance system in Japan, in which frequency and a period of physical therapy are restricted to 20 min per session and 2 days per week. We observed whether our intervention program prevented functional decline of the patient.

PARTICIPANT AND METHODS

In August 2016, a 41-year-old male diagnosed as HSP started using outpatient rehabilitation service in our Long-Term Care Health Facilities for the purpose of physical therapy. His chief complaint was difficulty in walking independently because of spastic paraplegia and blindness.

When he was in his late twenties, he felt difficulty in walking and decreased visual acuity. At the age of 32, he was diagnosed as HSP (causative gene was SPG 11: autosomal recessive mutation) in “A” hospital. Five years later, he underwent intrathecal baclofen therapy in “B” hospital but this therapy had no effect.

Spasticity and ataxia worsened progressively. He had been going outside without any walking aid or assistance until 2015. Since 2016, he used a wheel chair and needed the assistance of his friends to go outside. Additionally, he had walked for moving indoor, but there was an increase in the opportunity of creeping at home.

Sagittal section of T2 weighted MR image revealed strikingly thin corpus callosum and slightly atrophic cerebellar vermis (Fig. 1).

Visual acuity: hand-motion level.

Superficial and deep sensation: normal.

Tendon reflex: biceps (+/+), triceps (+/+), patella tendon (+/+), Achilles tendon (++/++), knee clonus (-/-), ankle clonus (+/+).

Muscle strength: MMT of the upper and lower extremities were both 4/5. Quadriceps (23.8/24.3 kgf), grip force (28.0/29.3 kgf).

Cerebellar function: Nose-mouth test (instead of finger-nose test), heel-knee test, and knee tap test were performed. Dysmetria (+), decomposition of movement (-), dysdiadochokinesis (+).

Intelligent function evaluated by WAIS (Wechsler Adult Intelligence Scale): verbal IQ was 71 (only verbal IQ was evaluated because of decreased visual acuity). For instance, WAIS values at 2012 were 71 in verbal IQ, 53 in performance IQ, and 58 in total IQ.

Gait analysis: The trunk was bent larger than normal in mid stance phase and in static standing position (Fig. 2). The posture at gait was wide-based because of truncal ataxia and spasticity of the lower extremities. He used Q-cane on the left hand. During the time period of mid stance phase and heel off phase of the gait, lateral bending and rotation of the trunk toward the side of the standing leg were observed with moderately larger degree than normal.

Home visit evaluation: He lived with a sister and father. His sister stayed mainly at home as a main caregiver, assisting preparation of meals, housework, and hospital visit. The ADL evaluated by the modified Barthel index was 65/100. Deduction items of the modified Barthel index were feeding, transfer ability, toilet, gait, stairs climbing, and dressing (Table 1). He mainly walked with Q-cane, but occasionally he crawled. He took a bath by himself once a week with great difficulty. Most of the time he lied on the bed. He scarcely went out except when he used outpatient service. His demand for the physical therapy was to prevent inability to walk alone. On the other hand, his family’s hope for the physical therapy was to maintain the ability of taking a bath alone.

Table 2 shows our original intervention program. The purpose of our intervention program was to maintain lower limb function and to keep the ability to take a bath alone. As we had to provide a rehabilitation program adapted to the limitations of long-term care insurance system in Japan, we set the frequency of rehabilitation substantially low compared with a conventional rehabilitation program. Practically, he underwent 40 min intervention for 2 days a week for 1 month. Then he continued 20 min intervention and 20 min personal practice (total 40 min practice) for 2 months. With reference to the degree of intensity, this program was planned according to the guidelines presented by American College of Sports Medicine (ACSM)¹⁰, except that the degree of intensity was increased gradually. Physical condition was evaluated 3 months after the intervention.

Additionally, we used two original techniques in order to control the hip abduction¹¹. First, by holding the elbows of the patient straighten, we prevented forward tilting of the trunk, hip flexion, and excessive activities of upper limbs during exercise. Second, we selected kneeling position in order to reduce the activity of hip abductor, thereby learning exact position of the trunk more easily. In the trunk control exercise (Fig. 3), the therapist supported his elbows to minimize the support of upper extremities and maximize the activities of trunk muscles and hip adductor.

Written informed consent was obtained from the patient regarding the publication of a case report with masked photographs. Additionally, this study has obtained approval from the research ethics committee of Aomori University of Health and Welfare (Approval number: 1731).

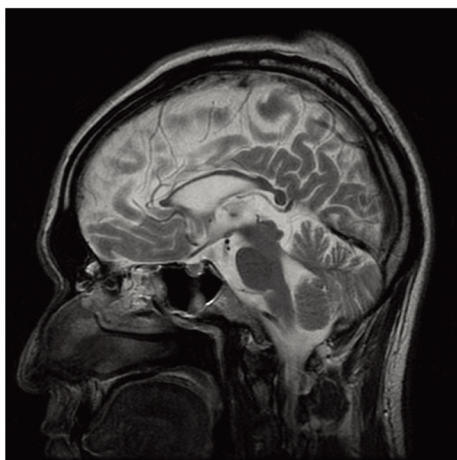


Fig. 1. Sagittal section of T2 weighted MR image. Corpus callosum is strikingly thin and cerebellar vermis is slightly atrophic.

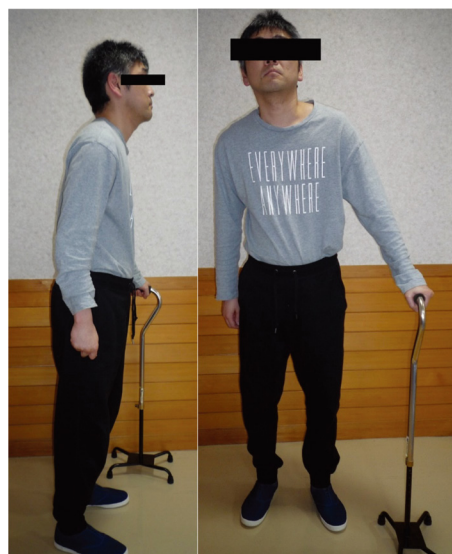


Fig. 2. Standing position of the patient using Q-cane on the left hand. Please note the wide-based stance and the trunk bent rightward.

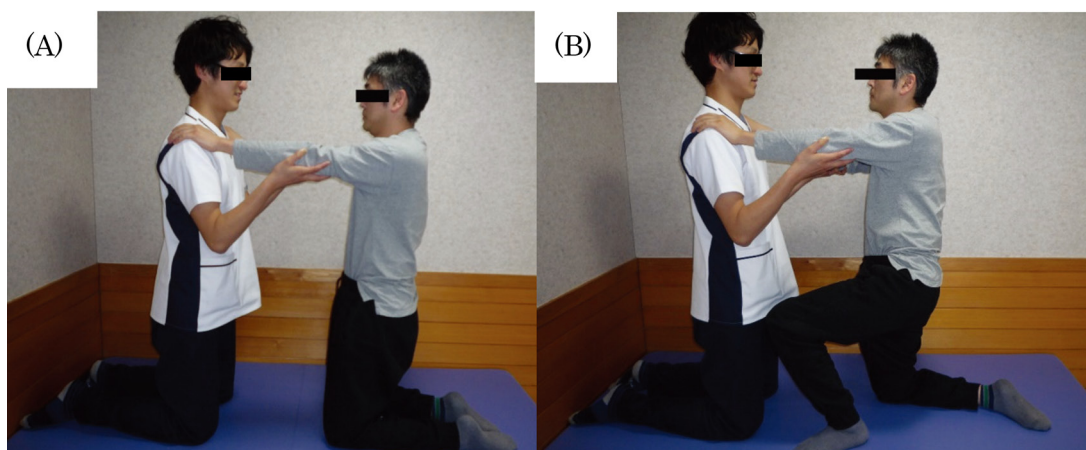


Fig. 3. Control exercise of hip abduction. (A) shows the starting position (kneeling position) and (B) shows the end position (half kneeling) of the exercise. The therapist supports the patients' elbows to hold them straighten.

Table 1. Original intervention program of physical therapy

No.	Intervention	Intensity
1	Squatting on the parallel bar	20 × 2 sets
2	Weight bearing exercise (kneeling gait sideways)	5 m × 10 sets
3	Control exercise of hip abduction (kneeling → half kneeling)	5 × 2 sets
4	Q-cane gait exercise	10 m
5	Motion exercise (taking a bath)	

Table 2. ADL evaluated by the modified Barthel Index

Items	Baseline	3 months
Feeding	5	5
Transfer ability	10	10
Personal hygiene	5	5
Toilet	5	5
Bath	5	5
Gait	10	10
Stairs climbing	0	0
Dressing	5	5
Bowel control	10	10
Bladder control	10	10
Total score	65	65

RESULTS

The patient underwent our original intervention program for 24 sessions (2 sessions a week × 4 weeks × 3 months). The number of repetition of squatting exercise was gradually increased to 100 times per session in the last half of the intervention.

At the beginning of the intervention, grip force was 28.0/29.3 kgf and quadriceps strength was 23.8/24.3 kgf. Three months after the intervention, grip force was 32.0/31.7 kgf and quadriceps strength was 26.3/28.0 kgf, indicating that the muscle strength was maintained throughout the intervention period. In addition, all scores of each item of Barthel index showed no change. He was still able to take a bath once a week and no accident happened in the bath at home. The posture at standing and gait showed no remarkable change.

DISCUSSION

We succeeded in maintaining the function of lower extremities and ADL abilities of the patient with complicated form of HSP by applying our original intervention program adapted to the limitations of Japanese long-term care insurance system in which time and frequency of rehabilitation is restricted.

Asir et al.⁹⁾ reported functional improvement of the patients with HSP by providing their original program in which frequency of the exercise was 6 sessions a week. In contrast to their program, we set the frequency of rehabilitation to 2 days a week, which amounted less than half of Asir's frequency. Without any exercise, functional decline should be predicted in a patient with HSP. However, our low exercise frequency program resulted in the maintenance of muscle strength, ADL abilities, and posture. These results suggest that low exercise frequency probably be enough for the maintenance of physical function in patients with HSP.

In order to keep the muscle strength, we considered the frequency and intensity of the training. ACSM's guideline indicates that strength training is needed at least 2 days per week to reinforce the muscle strength¹⁰⁾. In addition, the muscle performance at approximately 20–30% one repetition maximum level¹⁰⁾ is needed to maintain the muscle strength. The muscle activities of that level can be obtained during squatting exercise¹²⁾. Therefore we provided squatting exercise to the patient and succeeded in maintaining the muscle strength. As the patient had decreased visual acuity, we provided verbal instruction about trunk position and knee flexion angle. This verbal instruction might have also made a positive contribution to the maintenance of the muscle strength.

In order to maintain the posture at standing and gait, we paid attention to the activity of gluteus medius muscle and the position of hip joint while body weight was loaded. Spasticity of the hip adductor is one of the key features in HSP patients⁷⁾. In addition, he bent the trunk lager than normal in mid stance phase and in static standing position. The gluteus medius originates mainly from the iliac crest and ends at the greater trochanter¹³⁾ and works most effectively when the hip joint is at an extended position. In order to increase the activity of gluteus medius, we prevented the trunk tilting forward and prevented hip flexion by holding his elbow straighten. Consequently, the posture at standing and gait remained unchanged. Our method probably prevented the worsening of the posture. In addition, kneeling exercise also contributed to the improvement of weight control abilities of trunk and hip joint. However, kneeling exercise may have reduced the opportunity for unified control exercise using ankle, knee, and hip.

In addition, the motion exercise we performed in accordance with the environment of his bathroom at home was effective in the maintenance of ADL abilities.

Finally, by applying our original intervention program adapted to the limitations of long-term care insurance system in Japan, we were able to maintain the functions of lower extremities and ADL ability in a patient with HSP for at least 3 months. The present results seems to have the beneficial effect of low exercise frequency in the physical therapy of patients with HSP at outpatient rehabilitation service.

Conflict of interest

None.

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