Case Report

PHYSICAL THERAPY INTERVENTIONS FOR THE PATIENTS WITH HEREDITARY SPASTIC PARAPARESIS–AN EXPLORATORY CASE REPORTS

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ABSTRACT

Background and purpose: Hereditary spastic paraplegia (HSP) is a genetic disorder characterised by progressive weakness and spasticity of the lower limbs. To date, the structured rehabilitation programme for the patient with HSP is not documented. The objective of the study is to illustrate the effectiveness of the structured 8-week intensive rehabilitation programme (SEIRP). **Case description:** Two middle aged adult patients with HSP were included in this case reports. Both of them were brothers and sisters and classified under pure HSP type. Case 1 was a 45-year old with equinovarus deformity, paraparesis and severe spasticity. Case 2 was a 43-year old female presented with toe walking on left due to severe spasticity and hyperreflexia of the lower limbs. **Interventions:** The participants were given intensive physiotherapy programme which includes, stretching, strengthening and functional exercise program for the period of 8 weeks. Each treatment sessions lasted for about 60-90 minutes per day, 6 days per week. **Outcomes:** The outcome measures used were Timed Up and Go test (TUG), Functional Reach Test (FRT), timed 10 m Walk Test (10mWT) and Two-minute Walk Test (2MWT). Baseline measurements were taken before initiating the intervention, at 4th week and end of 8th week. **Results:** On completion of 8-week intensive physiotherapy programme results in Improvements were noted in all the outcome measures. **Conclusion:** SEIRP may be feasible and beneficial in improving the functional abilities in patients with HSP.

KEY WORDS: HEREDITARY SPASTIC PARAPLEGIA; REHABILITATION; INTENSIVE EXERCISE PROGRAMME; STRUCTURED PROTOCOL; PROGRESSIVE LOWER LIMB SPASTICITY.

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INTRODUCTION

Hereditary spastic paraparesis (HSP) or the Strümpell-Lorrain syndrome is the name given to the rare inherited autosomal dominant disorders in which the main clinical feature is progressive lower limb spasticity. From the epidemiological studies, the prevalence rate ranges between 4.3 and 9.6 per 100000 populations.[1,2] Features of HSP include slowly progressive spasticity, lower limb weakness and other pyramidal signs with little or without upper limb involvement. Harding (1983) classified the disease into two groups, one beginning before 35 years and other in their fourth to sixth decade of their life.[3] The former one shows protracted course and the later one shows sensory loss, urinary symptoms and action tremor.[4] Well planned rehabilitation programme prevents complications such as contractures and maximizes functional chores.[5] To our knowledge, till date there is no published literature describes the importance of rehabilitation in HSP. Hence, we intended to report a structured rehabilitation programme for the two cases with HSP.

CASE REPORT

Case no. 1: A 45-year old man, diagnosed as HSP was presented in our clinic for evaluation and rehabilitation. His chief compliant was difficulty in walking independently with frequent

history of falls. He was born of non-consanguineous marriage and the delivery was full-term and normal vaginal without any neonatal complications. During his perinatal period there were no unusual problems and he experienced no serious illnesses or injuries in his infancy and childhood. Based on family history, the individual mother, sister and brother are affected, the maternal uncle and grandmother are also affected (Figure 1).

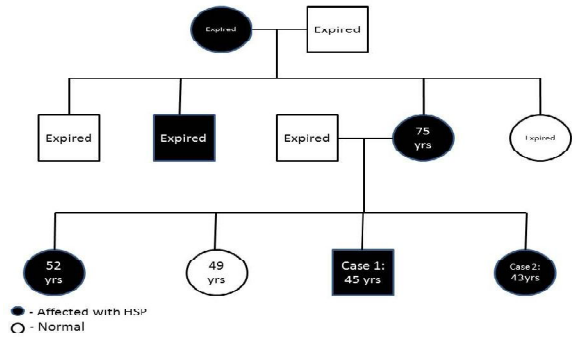

Figure 1: Pedigree of the reported family with HSP.

By the age 33, he developed slowly progressive symptoms. On examination he had a normal higher function. Examination of the lower extremity showed an exaggerated foot arches bilaterally with typical features of equinovarus deformity (Figure 2a), paraparesis (4/5 in lower limb), severe spasticity, tendon reflex were hyperactive with clonus (4+ DTR) of the both ankles, extensor plantar responses, decreased lower-extremity vibration and joint position sense. He had normal urinary sphincter functions and walked with crossing one foot over other using caregiver support. He is more concerned about his offspring bearing the same symptoms. So, he did not marry till date.

Figure 2: Case 1 (a), and case 2 (b) participated in the case series.

EEG, visual evoked responses, ophthalmoscopic exam, electromyography (EMG), and nerve conduction studies were normal. Spinal magnetic resonance imaging (MRI) showed a mild atrophy of the lumbar spine. MRI of the brain was normal.

Case no. 2: A 43-year-old woman (the first case's sister who was identified by us during our community based rehabilitation programme, one month later) was affected with the similar problems as his brother, but she developed the symptoms by the age 13. She was presented with frequent history of loss of balance with normal higher mental and cranial nerve function, non-fluent speech, bilateral genu recurvatum, equinovarus deformity and pes cavus with evident toe walking on left (Figure 2b). The video illustrating her gait can be viewed from http://youtu.be/B1oplU4dKhk. She presented a severe spasticity and hyperreflexia of the lower limbs with extensor plantar responses and ankle clonus. Vibration and joint passive movement sense at toes of both feet were reduced bilaterally. Muscle strength in the upper limb was normal and 4/5 in lower limb. Other paraclinical investigations were also normal. MRI of the brain is a normal study while spine revealed axonal degeneration of the terminal ends of corticospinal tract and posterior column in lumbosacral region.

PHYSIOTHERAPY INTERVENTIONS

The intensive rehabilitation programme was planned with the modification of guidelines suggested by the American College of Sports Medicine (ACSM),[6] targeting by improving the functional performance such as walking and balance. The details of the structured 8-week intensive rehabilitation programme (SEIRP) are tabulated in Table 1. Both the individuals participated in 8-weeks intensive programme, 6 days / week. Each sessions lasting about 60-90 minutes and one session / day, which includes regular warm-ups, stretching and strengthening. Their intensities were progressed gradually.

Assessments: Balance was assessed using FRT and TUG test, while walking ability by 2MWT and timed-10mWT. All the above mentioned outcomes were assessed before initiating the SEIRP, at 4th week and at the end of 8th week. Written

informed consent was obtained from both the individual regarding the publication of their case reports with masked photographs.

S.No.	Intervention	Intensity
1	Cycling	(8-15) min
2	Static Marching	33 steps x 3 reps
3	Stretching of hamstring, quadriceps, Iliopsoas, and TA	40 secs x 6 reps
4	Strengthening of quadriceps with 2-4 kg weight cuff and dorsiflexors with 0.5-2 kg weight cuff (on Monday, Wednesday and Friday), hamstring with 2-4 kg weights cuff , hip abductors and evertors with theraband (on Tuesday, Thursday and Saturday)	12 x 3
5	Heel rises on a wooden block	(20-30) rises x 3 reps
6	Step up on stepper	12 x 3
7	Squatting with therapy ball support on wall (Figure 4.a)	12 x 3
8	Sitting on therapy ball and performing sit-to-stand by attempting to get the ball held in therapist hand (Figure 4.b)	12 x 3
9	Sitting on therapy ball and catching the ball thrown to them	12 x 3
10	Catching and throwing the ball when performing sit-to-stand and stand-to-sit on therapy ball	12 x 3

Abbreviations: TA, Tendo Achilles; reps, repetitions; kg, kilograms.

Table 1: Structured eight week intensive rehabilitation programme for the individual with HSP.

RESULTS

Evident improvements were seen in their balance and walking ability during and at end of SEIRP from Figure 3 (a) (b) (c) and (d). The case 1 could able to walk independently for about 70.2-m and case 2, his sister could walk for about 25.4-m in 2 minutes. The SEIRP may be beneficial to rehabilitating the individual with type I HSP.

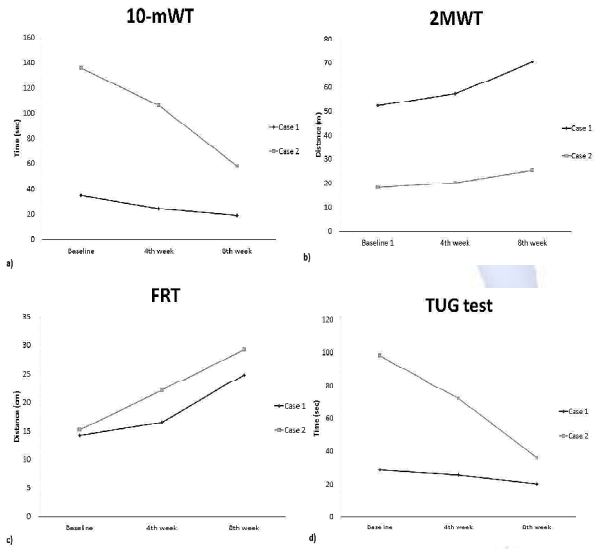

Figure 3: (a), (b), (c), and (d) Outcomes of balance and functional mobility in SEIRP.

DISCUSSION

This case series shows that the SEIRP improves functional ability of both the individual affected with HSP. They represent the large number of individuals to whom SERIP may be beneficial. Here their spasticity interferes in functional activities than their muscle weakness. Our first aim in rehabilitation is to minimize their spasticity to maximize their function. This was attempted by the slow rhythmic cycling as an initial intervention to improve lower limb muscle strength. Arousal was kindled by activating the reticular activating system, by stepping on heel towards the ground which sends signals to spino-reticular pathway.[7] We attempt to maintain the body erects by the above explained mechanism. Elongation of the shortened muscle and strengthening were done in accordance with ACSM guidelines.[6] 10 Repetition Maximum was not estimated, as we believed that spasticity might prevent in using their maximum force. Resistance was given according to their perceived tolerability. Conventional rehabilitation programmes were continued till the end of fourth week. Step up on the stepper, modified wall squatting on therapy ball (Figure 4a), stair climbing and sit-to-stand from therapy ball (Figure 4b), obstacle clearance (http://youtu.be/vakYGBa8CIY), stair climbing (http://youtu.be/ypF8SE8VYqc) and sit-to-stand from therapy ball with catching and throwing throw ball thrown by therapist (http://youtu.be/ymmTuTaPUrI) forms (S.No 6-11 in Table 1) the functional rehabilitation programme.[8]

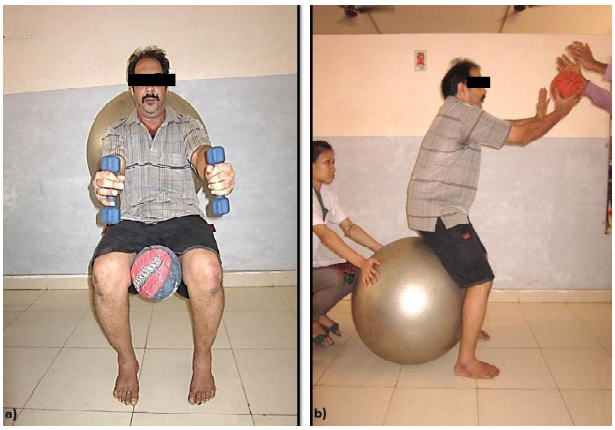

Figure 4: (a) Individual performing functional rehabilitation exercises. Modified wall squat with therapy ball, **(b)** Attempting to get the ball held in therapist hand.

We added the above functional rehabilitation exercises from fifth week to 8-week, the end of intervention. Both the individual with HSP, showed improvement in all the four outcome measures. But very significant improvements were noted in 10mWT and TUG from 5-8 week. Thus improving gait speed and decreasing the risk of fall. This may attribute to the functional rehabilitation exercises. As these exercises strengthen muscle force generation, maintain muscle length and improve intersegmental control of limb by improving neural control.[9] An individual to remain functional in day-to-day activities, they must possess these qualities. Moreover, these exercise programmes contain variety of experiences and activities to prevent boredom. We followed for only 8-weeks; however we believe greater improvements over long-term period.

The results of this case series should be interpreted with caution and there lies a need to verify the results with large number of subjects leading to randomized controlled trial (RCT).

CONCLUSION

These two cases highlight the importance of functional rehabilitation programme. It illustrates the functional improvement gained over the structured 8-week intervention programme in this population. The above findings suggest that a SEIRP may improve the functional level of the individual suffering from HSP.

Ethical approval: Not applicable. Written informed consent regarding the publication of individual data with masked photographs was obtained.

Conflict of interest: None declared. The authors alone responsible for treating the individual and writing of their report.

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