



**ORIGINAL RESEARCH PAPER**

**Physiotherapy**

**RESPONSE OF A HEREDITARY SPASTIC PARAPARESIS PATIENT TO CONSECUTIVE EIGHT WEEK REHABILITATION PROGRAM (CEREP) – A SINGLE CASE STUDY**

**KEY WORDS:** Hereditary Spastic Paraparesis, Consecutive Eight Week Rehabilitation Programme, Lower Limb Spasticity.

<b>Dr. Ankit Sinha</b>	Assistant Professor, Swarnim Start-up and Innovation University, Gandhinagar 1
<b>Dr. Suramya Shrama*</b>	Assistant Professor, USB college of Physiotherapy Aburoad. *Corresponding Author
<b>Priya kanjani</b>	Student, USB college of Physiotherapy, Aburoad
<b>Ravi Kumar Joshi</b>	Student, USB college of Physiotherapy, Aburoad

**ABSTRACT**

**BACKGROUND:** Hereditary spastic paraparesis(HSP) is a genetic disorder characterized by progressive weakness and spasticity of lower limbs. There has not been any single case study on intensive rehabilitation on hereditary spastic paraparesis patient. The objective of the study was to assess the response of a HSP patient to consecutive Eight Week Rehabilitation Program.

**CASE DESCRIPTION:** This case involved a 16 year old girl with spastic paraparesis that was 5-6 years in duration and slowly worsening. Her complaints mainly included difficulty in walking and keeping feet straight on ground since last 6 years. Both subjective and objective examination was conducted and neurological assessment was done. No bowel, bladder or any loss of sensory system was found.

**METHOD:** Balance was measured using Berg Balance Scale, 10 Meter Walk Test(10MWT) and Timed Up and Go(TUG) Test. Pre measurements were taken before the treatment began and in the end at 8<sup>th</sup> week. Data were computed using SPSS 17 software package.

**INTERVENTIONS:** This included stretching, strengthening, MAT Exercises and gait training for the period of 8 weeks. Each treatment lasted for 60-90 minutes per day, 5-6 days a week.

**RESULTS:** The result of the study is not quite a lot statistically significant, but clinically there is significant change in patient condition.

**CONCLUSION:** The findings of the study show that an 8 week rehabilitation program improves overall balance and gait patterns in patient.

**INTRODUCTION:**

HSP is also known as Hereditary Spastic Paraplegia, Familial Spastic Paraplegia, French Settlement disease or Strumpell - Lorrain disease. The symptoms are a result of dysfunction of long axons in the spinal cord. The affected cells are the primary motor neurons, therefore the disease is an upper motor neuron disease.<sup>1</sup>Worldwide, the prevalence of all Hereditary Spastic Paraplegias combined is estimated to be 2 to 6 in 100000 people.<sup>2</sup>Briefly, the clinical picture in pure HSP is of a slowly progressive, predominately symmetrical, spastic paraplegia. This is frequently accompanied by minor sensory abnormalities (such as absent vibration sensation) and neurological bladder involvement, but bowel involvement is rare.<sup>3</sup>In complex HSP, on addition wide range of neurological features including ataxia, extra pyramidal signs, epilepsy, mental retardation, dementia and peripheral nerve involvement occurs.<sup>4</sup> In pure HSP, life span is not affected.<sup>5</sup> People with HSP complain of muscle stiffness, pain, spasm and cramps, tripping over their toes due to weakness of ankle dorsiflexors and hip flexion, loss of balance, effortful walking and progressively more flexed standing posture<sup>3</sup>. Eventually, walking becomes impossible for some patients, due to a combination of (a) spasticity, (b) weakness particularly of ankle dorsiflexors, (c) loss of range of movement at ankle, knees and hip, making it impossible to stand straight, and (d) loss of motor control leading to delayed postural reflexes and loss of balance. Initial diagnosis of HSP relies upon family history, the presence or absence of additional signs and the exclusion of other non - genetic causes of spasticity.<sup>6</sup>A home exercise program supervised by a patient concentrating on stretches to maintain ROM and reduced spasticity, accomplished by balance exercises in patients with more advanced disease, is the cornerstone of management.<sup>7</sup>There is a recent literature supporting the use of intensive physical therapy i.e. stretching, strengthening, functional re-educative exercises about HSP and fatigue management<sup>2</sup>All these elements attracted our attention and curiosity about how patient with HSP perceive physical therapy and so we devised eight week rehabilitation programme.

**CASE REPORT AND ASSESSMENT:**

The patient is a 16 years old girl who began facing problems in walking at 10 years of age which was slowly worsening, showing cardinal features of hereditary spastic Paraparesis. She has developed scissoring type of gait pattern, has weakness of Gluteus Maximus muscle on the left side. Neurophysiological study shows UMN type of weakness in left lower limb. The case was diagnosed by consulting pediatrician 1 month ago and was referred for physical therapy intervention. She showed no familial history of inheritance or even related disorder. Subject was born out of normal vaginal delivery. Subject's mother has a history of frequent fever during the gestational period and even after the delivery. First symptom began to appear when patient was 10 years old. Spasticity was found in adductors group. Deformities like anterior pelvic tilt, internal femoral torsion, internal tibial torsion, genu valgum, pes planus and toeing out was present. Deep Tendon Reflex examination showed positive plantar reflex bilaterally. Electrophysiological testing of left lower limb showed UMN type of weakness.



**Figure 1: Foot arches, left foot signifying pes planus.**



**Figure 2: Gait pattern of the individual showing scissoring type of pattern.**

**ASSESSMENTS:** Balance was assessed using BBS and TUG, while walking ability by 10MWT . Pre and post measurements were taken before initiating the program and at the end of 8th week. Written informed consent was obtained regarding publication of their case reports.

**PHYSIOTHERAPY INTERVENTIONS:** Consecutive eight week rehabilitation program was designed to improve functional performance. Stretching, strengthening, gait training and MAT exercises were added. Duration of session was 60-90 minutes/day for 5-6 days a week which lasted for 8 weeks. Intensities were increased gradually based on performance.



**Figure 3: Individual performing MAT exercises.**

**TABLE 1: Consecutive eight week rehabilitation program for patient with HSP.**

INTERVENTION	INTENSITY
1) STRETCHING OF TA, ILIOPSOAS, ADDUCTORS, HAMSTRINGS	20 sec hold/ 4 reps
2) CORE MUSCLE TRAINING (supported bridging)	10 sec/ 10 reps
3) SPINAL MOBILIZATION	
4) STRENGTHENING EXERCISE (SLR, Gluteus medius with assistance and squatting)	10 reps
5) MAT EXERCISE (kneeling, half kneeling)	10 sec/ 10 reps
6) GAIT TRAINING (forward and sideway walking)	

**STATISTICAL ANALYSIS:**

After the collection of data according to BBS, 10 MWT and TUG, it was analyzed by SPSS 17 software package.

**Table 1:**

Descriptive Statistics					
	N	Minimum	Maximum	Mean	Std. Deviation
Prewtr	3	10.50	10.90	10.6667	.20817
Valid N (listwise)	3				

The mean value for 3 consecutive Pre 10 Meter Walk Test at regular speed was 10.6±0.20

**Table 2:**

	N	Minimum	Maximum	Mean	Std. Deviation
Pre 10MWT fast	3	9.60	9.80	9.7000	.10000
Valid N (listwise)	3				

The mean value for the 3 consecutive Pre 10 Meter Walk Test at fast speed was 9.7±0.10.

**Table 3:**

	N	Minimum	Maximum	Mean	Std. Deviation
Post 10MWT regular	3	9.70	9.80	9.7667	.05774
Valid N (listwise)	3				

The mean value for the 3 consecutive Post 10 Meter Walk Test at regular speed was 9.7±0.5.

**Table 4:**

	N	Minimum	Maximum	Mean	Std. Deviation
Post 10MWT fast	3	8.20	8.70	8.5000	.26458
Valid N (listwise)	3				

The mean value for the 3 consecutive Post 10 Meter Walk Test at fast speed is 8.5±0.2.

**Table 5:**

	N	Minimum	Maximum	Mean	Std. Deviation
Pre TUG	3	11.40	11.43	11.4133	.01528
Valid N (listwise)	3				

The mean value for the 3 consecutive Pre Timed Up and Go test is 11.41±0.1.

**Table 6:**

	N	Minimum	Maximum	Mean	Std. Deviation
Post TUG	3	8.60	8.80	8.7000	.10000
Valid N (listwise)	3				

The mean value for the 3 consecutive Post Timed Up and Go test is 8.7±0.1.

**DISCUSSION:**

The main aim of this study is to show that a course of 8 week rehabilitation program improves the functional ability of the patient affected with HSP.

The evaluation of walking speed is widely used in physiotherapy assessment for patient with neurologic diseases (according to Molteni F and et. al). A gait speed of less than 1m/s identifies a person at high risk for negative health outcomes. As well as a study done by Mark Braschinnskyet.al.in 2009 showed that person with HSP represent a high risk group for the 10 min walk test and walking speed in the HSP was more influenced by ROM of hip muscle. In our study we used 3 main outcome measures namely: 10 meter walk test, Berg Balance Scale and Timed Up and Go Test. The values over 8 week didn't show that much statistical improvement but clinically the walking speed and balance was improved. The possible reason for this could be that due to some personal reason she wasn't able to follow up, despite this the condition didn't deteriorate. The possible reason being that she was following the home program being given to her. There were significant improvement in Range Of Motion and Manual Muscle Strength Testing following CEREP. A previous study done by Mark et. al. showed that limited Active ROM and increase spasticity on Modified Ashworth Scale results in reduction in walking speed. So this support our results, walking was improved as spasticity was reduced, AROM increased along the course of time. (voluntary muscle grading (VGC) was more than 4).

These results would have been statistically significant if the treatment would have been regular for whole 8 weeks. Patient was also taking intervention prior to the initiation of this study which could also be a reason for marked improvement in gait, balance, ROM and MAS. The results of this study should be interpreted carefully, and it should be verified over larger patient population.

**CONCLUSION**

The findings of the study show that an 8 week rehabilitation program improves overall balance and gait patterns in patient. Clinically improvements were seen but not relevant statistically. The improvement confirms the therapeutic benefits and benefits of home program both.

**LIMITATIONS AND RECOMMENDATIONS:** Although the data analysis revealed significant findings to support our result, there are still limitations in this study.

**LIMITATIONS:**

- 1) Short duration of study as patient was not able to follow up till the end
- 2) Limited sample
- 3) Aspects of cardiac endurance were not tested which are often associated with impaired gait.

**RECOMMENDATIONS:**

- 1) Long term effect should be seen
- 2) Study over larger population can be done
- 3) Fatigue management technique can be incorporated
- 4) Comparative study can be done with other neurological disease.

**REFERENCES:**

- 1) Depienne, Christel; Stevanin, Giovanni; Brice, Alexis (2007-12-01). "Hereditary Spastic Paraplegias an update." *Current Opinion in Neurology* :20 (6):674-680
- 2) National Institute of Health (2008). "Hereditary Spastic Paraplegia Information Page." Retrieved 2008-04-30.
- 3) AnkeHensiek, Stephen Kirker and Evan Raid "Diagnosis, intervention and management of hereditary spastic paraplegias in the era of next-generation sequencing." *JNeurol*, 2015; 262(7): 1601-1612
- 4) Salinas S, Proukakis C, Crosby A, Warner TJ. Hereditary Spastic Paraplegia: clinical features and pathogenetic mechanisms. *Lancet Neurol* 2008; 7(12): 1127-38.
- 5) Fink Jk. Hereditary Spastic Paraplegia overview. 2000 Aug 15 [updated 2009 Feb 03]. In: Pagon RA, Bird TD, GeneReviews. Seattle: University of Washington
- 6) Harding AE, Classification of the hereditary ataxias and paraplegias. *Lancet* 1983; 1: 1151-1155
- 7) Geva - Daupan K, Domenievitz D, Zahalka R, FattalValenski A. Botulinum toxin injections for pediatric patients with hereditary spastic paraparesis. *J. Child Neurol* 2010; 25:969-975
- 8) Asir John Samuel, VenticaPriyankaAranha, TrapthiKamath. Physical Therapy Interventions for the patients with Hereditary Spastic Paraparesis- An Exploratory Case Reports; *IJPR* 2013 (3): 110-13