Spinal Cord (2007) 45, 64–68

www.nature.com/sc

Original Article

Disability and determinants of gait performance in tropical spastic paraparesis/HTLV-I associated myelopathy (HAM/TSP)

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Study design: Cross-sectional.
Objectives: The aim of this survey is to describe the disability profile in a group of tropical spastic paraparesis/HTLV-I-associated myelopathy patients, identifying the requirements for community ambulation.
Setting: Terciary care unit, Rio de Janeiro, Brazil.
Methods: Seventy-two patients were assessed (49 female and 23 male), referred by tertiary care centers, when a clinical protocol was applied.
Results: The sample had an average age of 40 years and an average of 137 months of duration of the disease. The most prevalent aspects of disability found were in gait and sphincter control areas. A total of 72% of the patients were community ambulators and 17% were restricted to wheel chair. Age, strength and low-back pain interfere in activities of daily living ($P<0.05$). A positive correlation was found between community ambulation and the knee extensors ($r=0.80$) and ankle plantar flexors ($r=0.74$). Strength, age, low-back pain, duration of disease, asymmetric onset of the symptoms and spasticity interfered in the ability to walk ($P<0.05$). A rehabilitation program was proposed focusing on modifiable factors that affect disability level.
Conclusion: It was possible to describe the profile of disability in this group of patients, identifying the requirements to the community ambulation.
Sponsorship: Not applicable.
Spinal Cord (2007) **45,** 64–68. doi:10.1038/sj.sc.3101919; published online 28 March 2006

Keywords: HTLV-I; disability; myelopathy

Introduction

Tropical spastic paraparesis (TSP) is a myelopathy clinically characterized by a chronic and progressive spastic paraparesis, related to several degrees of sphincter and sensory disturbances, being predominant in tropical areas.[1] In 1985, Gessain *et al*[2] showed its relation with the human T-lymphotropic virus type I (HTLV-I). A similar neurological condition was described soon after in Japan and was named HTLV-I-associated myelopathy (HAM).[3] In 1988, both were recognized as being the same disease. The term HTLV-I-associated myelopathy/tropical spastic paraparesis (HAM/TSP) has been adopted ever since.[4] It is estimated that approximately 20 million individuals are infected worldwide.[5] The lifetime risk for developing HAM/TSP in asymptomatic carriers is between 1 and 5%.[6]

The predominant clinical feature of HAM/TSP is a spastic paraparesis with sphincter disturbances. The neurological examination generally shows weakness and spasticity in the lower limbs with an adductor pattern.[7,8] Because of its high prevalence in Brazil, there has been an increasing demand for rehabilitation treatment of individuals with HAM/TSP in the last years. This study aims at establishing the major characteristics of disability in subjects with HAM/TSP and to identify variables related to community ambulation.

Methods

The initial sample involved 82 subjects consecutively referred to the Clementino Fraga Filho University

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Hospital (HUCFF) Physical and Rehabilitation Service of the Federal University of Rio de Janeiro, from two reference centers for this disease in the city of Rio de Janeiro, Brazil. The exclusion criteria were other concomitant neurological diseases, coinfection with HIV, diabetes and alcoholism (owing to the high prevalence of polyneuropathy in these diseases) and orthopedic diseases. Eight subjects were excluded from the sample because cerebrospinal fluid results were not available and two for having knee and hip prosthesis. The remaining 72 patients fulfilled the diagnostic guidelines of the World Health Organization for HAM/TSP.[9] The study was submitted and approved by the Ethical Committee of HUCFF, and all subjects signed an informed consent.

The subjects were assessed by the same physiatrist, in one visit, when the clinical disability protocol was fulfilled. None of these individuals were in a formal rehabilitation program.

The protocol included history, neurological and physiatrist examination. The ambulation functional classification[10] and the use of walking aids were specified. Spasticity was assessed using the Ashworth scale.[11] The presence of action hypertonia, categorized as mild, moderate or severe was observed during gait. The presence of contractures was assessed in hip flexors, hip adductors, knee flexors and ankle extensors using Kendall's maneuvers.[12] The Manual Muscle Test (MMT)[13] was performed bilaterally in hip flexion, hip abduction, hip extension, knee extension, knee flexion, ankle flexion, ankle extension and hallux extension.

The ambulatory motor index (AMI)[14] and the Lower Extremities Motor Scores of the American Spinal Injury Association – ASIA-LEMS[15] were used. The sensory examination included pin prick and light touch sensation, according to the International Standard Classification of Spinal Cord Injury,[15] and joint position sense.

The functional independence measure (FIM)[16] was used as the daily living activity scale.

The sample was then divided according to their ambulation functional levels into

- group 1: subjects were community ambulators;
- group 2: subjects were household ambulators or non-ambulators (wheelchair-bound).

The groups were compared in terms of muscle power, FIM, age at onset of disease, age at assessment, duration of the disease, presence of low-back pain, asymmetric onset of symptoms and spasticity. Correlation coefficients were used to determine the association between the studied variables. The student's *T*-test was used to measure significant differences between these groups ($P<0.05$).

Results

The sample included 72 subjects (49 female, 23 male). The mean age at onset was 40 years (SD 12.1), at assessment was 51.1 (SD 12.3) and the mean duration of the disease was 137 months (SD 83.7). History data are shown in Table 1.

Fifty two patients were community ambulators, 20 being household ambulators or wheelchair bound. The physical examination results are displayed in Tables 2 and 3.

The average muscular power indexes used were AMI 58% (0–100%) (SD 25%) and ASIA-LEMS 28 (0–50) (SD 10). The relation between walking aids and the means of muscular power indexes is shown in Table 4.

All subjects obtained maximum scores in the FIM cognitive items. The average FIM score was 108.

The strongest correlation between ambulation functional levels and muscle strength occurred in knee extension (0.80) and plantar flexion (0.74).

There was a highly positive correlation between right and left muscle strength ($\geqslant 0.93$). There was a negative although weak correlation (-0.12) between ambulation functional levels and action hypertonia. The correlation

Table 1 History data

	Yes	*No*	*Not known*
Asymmetric onset of symptoms	46 (63.9%)	26 (36.1%)	0
Gait disturbance	72 (100%)	0	0
Sphincter disturbance	71 (98.6%)	1 (1.4%)	0
Erectile dysfunction[a]	22 (95.7%)	1 (4.3%)	0
Low-back pain	47 (65.3%)	25 (34.7%)	0
Leg pain	24 (33.3%)	48 (66.7%)	0
Objective sensory abnormality	5 (6.9%)	67 (93.1%)	0
Using antiespastic drugs	61 (84.7%)	11 (15.3%)	0
Using orthosis	53 (73.6%)	19 (26.4%)	0
History of previous neurological illness	2[b] (2.8%)	70 (97.2%)	0
Family history of paraparesis	8 (11.1%)	63 (87.5%)	1 (1.4%)
History of breast feeding	61 (84.7%)	3 (4.2%)	8 (11.1%)
History of blood transfusion	21 (29.2%)	51 (70.8%)	0
History of drug abuse	10 (13.9%)	62 (86.1%)	0
History of sexually transmitted disease	26 (36.1%)	46 (63.9%)	0

[a]Male population ($n=23$)
[b]Epilepsy

between ambulation functional levels and Ashworth scale scores was positive but also weak (+0.14).

The correlation between muscle power indexes and ambulation functional level was positive (better muscle scores implying a better functional level), the ASIA-LEMS correlation being stronger (+0.7591) than AMI (+0.6423).

Table 2 Ambulation functional levels and walking aids

	n
Functional levels	
Community ambulators	52 (72.2%)
Household ambulators	8 (11.1%)
Wheelchair bound	12 (16.7%)
Walking aids	
None	19
Crutches	37
Walker	4
Wheelchair	19

Seven subjects used more than one type of walking aids

Table 3 Muscle contractures and sensory examination

	Normal	*Abnormal*
Muscle contractures		
Adductors	12 (16.7%)	60 (83.3%)
Plantar flexors	18 (25%)	54 (75%)
Hip flexors	24 (33.3%)	48 (66.7%)
Knee flexors	28 (38.9%)	44 (61.1%)
Sensory examination		
Light touch	66 (1.7%)	6 (8.3%)
Pinprick	63 (87.5%)	9 (12.5%)
Position sense	69 (95.8%)	3 (4.2%)
Sensory level*	69 (95.8%)	3 (4.2%)

*Levels: T3, T6 and T11

Table 4 Walking aids and muscular power indexes

	n	*AMI*	*ASIA-LEMS*
Without orthosis	19	84%	37
Crutches	32	58%	29
Walker	2	51%	26
Wheelchair + crutches or walker	7	41%	23
Wheelchair	12	30%	13

AMI: ambulatory motor index

Groups were compared in terms of

- muscle power,
- FIM,
- age at onset of disease,
- age at assessment,
- duration of disease,
- asymmetry of symptoms at onset of disease,
- presence of low-back pain,
- spasticity.

The results are shown in Tables 5 and 6.

Discussion

The epidemiological characteristics of the studied sample do not differ from what has been described in literature.[7,8] Patients were predominantly women (2:1) with an average age at onset of 40 years. Some subjects found it difficult to determine precisely the age at the onset of their disease, mostly because of the long period of the disease and the subtle and progressive installation of the initial symptoms. This fact has also been described in other series.[17,18] A total of 64% of the sample described an asymmetric onset of motor symptoms and 65% complained of low-back pain. Gait and sphincter disturbances were the most common complaints. The majority of individuals had community ambulation, with varying levels of difficulty and speed. The gait disturbance was also severe, because 74% of subjects needed walking aids. On the other hand, only 17% of patients were restricted to a wheelchair. These data disagree with Montgomery's report[19] in which after 10 years of the disease, 50% of subjects would be wheelchair bound. When the sample was analyzed according to the duration of the disease, from the group

Table 6 Comparison between functional groups

	Group 1	*Group 2*	P-*value*
FIM*	113	92	<0.01
Age at onset	38 years	44 years	0.07
Age at assessment*	48 years	58 years	<0.01
Duration of disease*	123 months	174 months	<0.02
Initial asymmetry*	60%	75%	<0.01
Presence of low-back pain*	59%	80%	<0.01
Ashworth 3 and 4*	10%	40%	<0.01

FIM: functional independence measure
*Statistically significant

Table 5 Muscular power indexes among functional groups

Indexes	*Group 1*	*CI*	*Group 2*	*CI*	P-*value*
AMI	68%	74–62%	31%	36–27%	<0.01
ASIA-LEMS	33	35–31	15	18–11	<0.01

AMI: ambulatory motor index; CI: confidence interval
Group 1: community ambulators; group 2: household ambulators and wheelchair bound

with less than 10 years of disease ($n=31$), 10% were restricted to wheelchair, and from the group with more than 10 years of disease ($n=41$), 22% were wheelchair bound. These data are similar to those of Nakagawa *et al*,[20] Araujo *et al*[21] and Cartier *et al*.[22]

The presence of muscle contractures complies with the gait pattern, the adductors being the most frequent involved muscles (83%). There were a minority of subjects with abnormal sensory examination (14%), in whom a distal deficit prevailed.

The correlation of muscle power and the ambulation functional classification showed that more strength entails a better ambulation. Two muscle groups showed stronger correlation with ambulation: quadriceps (0.80) and plantar flexors (0.74). The correlation between quadriceps muscle power and ambulation showed that among subjects with strength 0, 1 and 2, only one patient had community ambulation. This patient had a severe action hypertonia that could be helping him to walk. Of the 10 subjects with quadriceps muscle strength of 3, five had community ambulation. All 46 individuals with quadriceps muscle strength of 4 had community ambulation. The importance of the quadriceps has been described by other authors. It is considered essential for functional ambulation of traumatic spinal cord injury patients.[23,24] The importance of plantar flexor muscles, however, has been minimized.[25] The role of this muscle group in the gait cycle, being active in weight acceptance (loading response and pre-swing phase), is important to progress and stability. The heel rising in the beginning of the terminal stance demands ankle stability maintained by the plantar flexors; in the pre-swing phase, plantar flexor muscles help knee flexion.[26]

The correlation coefficient between right and left lower limbs was significant (>0.93). A positive correlation was also found when each movement was compared (0.63–1). Thus, it can be inferred that progression of power loss occurred in a global and uniform manner in the studied muscle groups.

The correlation coefficient between the ambulation functional classification and the muscular power index shows that the better the index, the better the ambulation. The AMI had a correlation of 0.64 and the ASIA-LEMS 0.76. The strongest correlation of ASIA-LEMS can be explained by the fact that it includes plantar flexion, whereas AMI excludes ankle and foot movements. As pointed out before, the muscle groups that are more interconnected with community ambulation are quadriceps and plantar flexors, both contemplated by the ASIA-LEMS. The muscular power index means and its confidence intervals did not differ from what is reported in traumatic spinal cord injury.[14] It is necessary to score above 30 in ASIA-LEMS to perform community ambulation.

The use of walking aids is inversely correlated with muscle power index scores. This fact has already been described in relation to AMI.[27] Thus, the indexes could be used to assist in prescribing suitable devices.

When divided into two groups according to strength – weak group (muscle strength 0, 1) and strong group (muscle strength 3 and 4) – the length of the disease seems to influence loss of muscle power in most of the studied movements (11 in 16); the disease was longer in the weaker group. This fact is also observed when functional groups and time of disease are compared; the group with greater disability in walk had a disease of longer duration.

The plantar flexors and knee extensors were the muscles that remained stronger for longer periods of time. Hip muscles (abductor, extensor and flexor) were the ones that remained stronger for a shorter period of time. This fact confirms the description of a proximal pattern loss of muscle power.[28] The slow development of disability observed in some individuals can be explained by the fact that the muscle groups that remained stronger for longer periods of time were those that have stronger correlations with community ambulation.

The Ashworth scale is commonly used as a clinical tool to assess spasticity.[29–31] Higher scores in the Ashworth scale were found in patients within the functional group of household ambulators and non-ambulators. The pattern of muscle activation and the increase of tonus in subjects with spasticity are different during movement and on passive mobilization.[32] Because the use of the Ashworth scale is not sufficient to assess spasticity in subjects with HAM/TSP, action hypertonia measurements are also necessary.

Age can be considered an additional factor for disability, as the group 2 of household ambulators and non-ambulators had a higher average age.

Low-back pain had a higher prevalence in group 2 (75%). It also seems to be related to the length of the disease. Within the subject group with less than 10 years of disease ($n=31$), 52% reported low-back pain, whereas 76% of subjects with more than 10 years ($n=41$) had this complaint. Pain can be linked to a lower level of activity, leading to restriction of mobility. Because pain is such a prevalent symptom, future studies are necessary to understand its mechanism so as to improve its treatment.

The performance of HAM/TSP patients in activities of daily living has already been published.[33]

The definition of a profile of disability in subjects with HAM/TSP allows a rehabilitation program prescription with more focused aims. The disease leads to various levels of disability, ranging from subjects with community ambulation to wheelchair-bound individuals. The ideal rehabilitation treatment must include a complete multidisciplinary team. Assessment of muscle strength should include motor indexes, preferably ASIA-LEMS, as it had a stronger correlation with the ambulation functional level. The indexes can also contribute to the prescription of walking aids. Strengthening exercises must be global, but focusing on the plantar flexors and quadriceps, muscle groups that are strongly correlated with community ambulation. The range of motion exercises should include the hip adductors and flexors, the knee flexors and the plantar flexors that are frequently shortened because of the gait pattern.

The aims of rehabilitation programs most focus on modifiable factors that might interfere with disability. Future research must be done to evaluate the effectiveness of the proposed treatment.

Conclusions

The profile of disability in a group of subjects with HAM/TSP was described and variables related to community ambulation identified. The ambulation functional level is related to the studied muscle groups, particularly the knee extensors and plantar flexors. These muscles should be focused in a program of strengthening exercises. The loss of muscle strength occurred homogeneously in this group of subjects. The ASIA-LEMS index presented the strongest correlation with ambulation functional levels. There was a weak correlation between ambulation functional levels and spasticity measurements. Ambulation functional levels are related to muscle power indexes, age of assessment, initial symptom asymmetry, presence of low-back pain and spasticity measured using the Ashworth scale.

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