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Original Article

Disability profile of patients with HTLV-I-associated myelopathy/tropical spastic paraparesis using the Functional Independence Measure (FIM™)

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Study design: Survey.
Objective: To determine the disability profile of a group of patients with human T-cell lymphotropic virus type I-associated myelopathy/tropical spastic paraparesis (HAM/TSP), using the Functional Independence Measure (FIM™) to identify the most affected functional areas.
Setting: Reference center for HTLV Rio de Janeiro, Brazil.
Methods: A total of 72 patients (49 female and 23 male), consecutively referred by tertiary care centers, were assessed using the FIM™.
Results: The average FIM™ score was 108 ($\pm$12 SD) ranging from 58 to 122. The lowest items scores were obtained in locomotion and bladder management. When divided into two groups (above, and below or equal to the average score), there were significant differences ($P<0.05$) in age at time of assessment, in the degree of muscular power and in low back pain. There were no significant differences in terms of age of onset and duration of the disease.
Conclusions: The most affected areas in FIM™ motor items were locomotion (walk and stairs) and bladder management. Age, strength in lower limbs and low back pain interfere with functional activities in patients with HAM/TSP. The duration of the disease is not a significant factor for patient disabilities. The goals of rehabilitation in HAM/TSP patients should target the modifiable factors, such as pain, strength and the neurogenic bladder.
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Introduction

The human T-cell lymphotropic virus type I (HTLV-I) is a retrovirus etiologically associated with HTLV-I-associated myelopathy/tropical spastic paraparesis (HAM/TSP).[1,2] HTLV-I can be transmitted by sexual contact with an infected individual, through sharing of contaminated needles and syringes by intravenous drug users, following transfusion of contaminated blood and from mother to child, through perinatal exposure or breast-feeding. The real prevalence of HTLV-I is still unknown, but it is estimated that 10 to 20 million individuals carry the virus worldwide. HTLV-I is endemic in many geographic areas including Japan, the Caribbean, Africa, South and North America and Melanesia. Infection rates vary widely in different geographic areas, ranging from less than 1% in certain European countries to as high as 30% in Southern Japan.

The real mechanisms of HTLV-I-induced diseases like HAM/TSP remain unknown. Although HTLV-I persists notwithstanding a strong immune response, only 2–3% of infected individuals will develop HAM/TSP. Most infected individuals are clinically asymptomatic carriers.[3–6]

The seroprevalence increases with age and is twice as high in females. HAM/TSP is a myelopathy characterized anatomopathologically by a chronic, progressive, low-grade inflammatory process heralded by parenchymal infiltration of memory CD4 cells. The inflammation involves both the gray and white matter of the spinal cord. Both the inflammation and the white matter degeneration are most conspicuous in the lower thoracic cord. The lateral funiculus is always and most severely affected. Although the parenchymal tissue degeneration

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is not confined to any particular long tract, symmetric degeneration of the lateral pyramidal tract is evident in all cases.[7]

The main clinical manifestation of HAM/TSP is a spastic paraparesis or paraplegia, characterized by a slowly progressive course of upper motor neuron involvement and mild sensory signs, coupled with sphincter disturbances. HAM/TSP is a disease with a slow onset and a chronic and steady progression. However, occasionally it can show a rapid deterioration.[8] Urgency, incontinence and difficulty in micturition are major complaints. Both irritative and obstructive urinary symptoms coexist in HAM/TSP patients.[9] Low back pain is a frequent complaint in HAM/TSP patients. However, there are no specific studies available regarding this symptom. It seems to be multifactorial and may exhibit musculoskeletal and neuropathic characteristics. The cognitive deficits described in association with HTLV-I are characterized by mild impairments in verbal and visual memory, attention and visual-motor abilities.[10] The progression of the disease is variable but often unremitting. Usually, the evolution is chronic, with progression over a number of years, finally reaching a plateau.[4,11]

HAM/TSP can be still associated to other HTLV-I-related manifestations such as pulmonary alveolitis, uveitis, arthritis, dermatitis, Sjögren's syndrome, Behçet's disease, thyroid disease, crusted scabies and cystitis and prostatitis.[12]

The Functional Independence Measure™ (FIM)[13] assesses physical and cognitive disability. It is used to monitor patient progress and to assess outcomes of rehabilitation.[14] Ratings consider performance rather than capacity and may be based on observation, a patient interview or medical records.[15] The FIM includes 18 items covering independence in self-care, sphincter control, transfers, locomotion, communication and social cognition. The seven-point rating represents gradations of independence and reflects the amount of assistance a patient requires. Scores range from a low of 18 to a maximum of 126.[13] Intraclass correlations, for pairs of clinicians rating 263 patients, range from 0.93 (locomotion subscale) to 0.96 (self-care and mobility). The mean kappa index of agreement between ratings for each item was 0.71.[16] The FIM™ is widely used for all aspects of disabling diseases including spinal cord injury,[17–19] allowing comparative studies among HAM/TSP patients and other disability groups.

In clinical practice, there is a high prevalence of complaints regarding gait and bladder management areas, in contrast to low prevalence of complaints regarding other activities of daily living (ADLs). Notwithstanding its high prevalence, we are not aware of studies on rehabilitation in HAM/TSP patients. The aim of this study was to identify the most affected functional areas in these patients and to compare the least and most dependent subjects in the sample so as to explore differences between them.

Methods

The study was submitted and approved by the Ethical Committee of Clementino Fraga Filho University Hospital (HUCFF) and all patients signed an informed consent. FIM™ was applied by direct interview of 72 HAM/TSP patients (49 female and 23 male) referred consecutively to the 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 (IPEC/HUCFF). The interviews were made by the same physiatrist (ACF). In the L item (Locomotion: walk or wheelchair), the variable scored was walk.

Patients were included in the survey if the WHO's HAM/TSP diagnosis guidelines[20] were met.

The exclusion criteria were

- other neurological diseases,
- HIV coinfection,
- diabetes and alcoholism,
- orthopedic diseases.

The initial sample had 82 patients. Eight patients were excluded because the cerebral–spinal fluid (CSF) results – an absolute requisite for diagnosis – were not available. The other two were excluded because they had hip and knee prosthesis. A clinical protocol with history, neurological and physiatrist examination was performed. The muscular power indexes employed were the Ambulatory Motor Index (AMI)[21] and the Lower Extremities Motor Scores of the American Spinal Injury Association – ASIA LEMS.[22]

The AMI is derived from manual muscle testing of five lower extremity muscles about the hip and knee. Muscle groups about the ankle are not represented. Bilateral motor scores for hip flexion, hip abduction, hip extension, knee extension and flexion are assessed. Grades 1 and 2 on manual muscle test score 1 in AMI, grade 3 scores 2, and grades 4 and 5 score 3. The sum of these scores was expressed as a percentage of the maximum possible score (30 points).

The ASIA LEMS is derived from manual muscle testing of five lower extremity muscle groups, representing each neurological level from L2 to S1 (hip flexors,

Table 1 Characteristics of the sample ($n = 72$ patients)

	Average	*SD*	*Range*	*%*
Age of onset	40 years	12.1	9–65 years	—
Age of assessment	51.1 years	12.3	17–78 years	—
Duration of disease	137 months	83.7	12–420 months	—
AMI (%)	58	25	0–100	—
ASIA-LEMS	28	10	0–50	—
Presence of low back pain	—	—	—	65%

knee extensors, ankle dorsiflexors, long toe extensors, ankle plantar flexors). Range of score: 0–50. Patient characteristics are described in Table 1.

The whole sample was studied for level of functioning in FIM™ ($n=72$). The sample was then divided into two groups, according to the average FIM™ score (group $1 \leqslant 108$ and group $2 > 108$), to compare the least and most dependent subjects, so as to explore differences between them.

The variables investigated were as follows: age at onset of disease, age at assessment, duration of the disease, strength and the presence or absence of low back pain; Student's *t*-test was employed. An α value $\leqslant 0.05$ was considered significant.

Results

The lowest levels of functioning in FIM™ motor items scores were observed in locomotion and bladder management. In all, 65% of subjects were independent in walk and stairs and 38% in bladder management. In all other motor items, at least 85% of subjects were independent. All subjects scored 7 on all five cognition items. The average FIM™ score was 108, ranging from 58 to 122. FIM™ motor items scores are shown in Table 2.

In the studied sample, 12 subjects were wheelchair restricted, and all scored 1 in walking. Of these patients, 66.7% would have scored 6 if the chosen variable had been wheelchair.

After the sample was divided into two groups, 22 patients scored equal or below average FIM™ scores (group $1 \leqslant 108$) and 50 patients scored above average FIM™ scores (group $2 > 108$). The variables investigated were as follows: age at onset of disease, age at assessment, duration of the disease, strength and the presence or absence of low back pain.

The sample average AMI was 58% (SD 25%); average ASIA-LEMS was 28 (SD 10). When divided into two groups according to the average FIM™, the most dependent group (FIM™ $\leqslant 108$; group 1) scored AMI = 36% and ASIA-LEMS = 18. The other group (FIM™ > 108; group 2) scored AMI = 68% and ASIA-LEMS = 33. The differences were significant, *P*-value = 0.001.

Low back pain is prevalent in 65% of subjects studied ($n=72$). Low back pain seems to be related to duration of the disease; 76% ($n=41$) of patients afflicted over 10 years complain of low back pain in contrast to only 52% ($n=31$) of patients afflicted under 10 years.

The variables that presented significant differences were age at assessment, presence of low back pain and strength in lower limbs. The results of the Student's *t*-test are shown in Table 3.

Discussion

The most affected functional area was bladder management. Only 38% of patients were independent. These low scores can be explained by the high prevalence of bladder sphincter disturbances[23–26] and also because the FIM™ scale considers state of continence in addition to independence in the management of neurogenic bladder.

Locomotion (walk and stairs) was the second most affected area. The chief complaint of 60 patients (83%) was gait disturbances, which was also the most prevalent symptom.

In other functional areas, such as self-care and mobility (transfers), patients were somewhat independent. This may be explained by the fact that motor symptoms are usually confined to legs, in spite of occasional involvement of arms.[27] Moreover, most patients presented paraparesis; few were paraplegics.

Table 3 Main differences between groups 1 and 2

	Group 1 (n = *22*)	*Group 2* (n = *50*)	P-*values*
Age at onset* (years)	43.3	38.6	0.130
Age at assessment* (years)	56.6	48.9	0.013
Duration of disease* (months)	159.7	128	0.140
Low back pain (% present)	77%	60%	0.001
AMI*	36	68	0.001
ASIA-LEMS*	18	33	0.001

Group 1: FIM $\leqslant 108$; Group 2: FIM > 108
*Group average

Table 2 Level of functioning in FIM motor items ($n=72$)

Level	*Eating*	*Grooming*	*Bathing*	*Dressing upper body*	*Dressing lower body*	*Toileting*	*Bladder management*	*Bowel management*	*Transfer–bed, chair, wheelchair*	*Transfer Toilet*	*Transfer tub, Shower*	*Walk*	*Stairs*
7	71	18	7	13	0	14	5	18	3	4	4	0	0
6	1	54	59	56	65	52	22	45	61	59	57	47	47
5	0	0	0	1	0	0	18	5	0	0	0	13	2
4	0	0	2	0	1	0	2	1	1	2	2	0	0
3	0	0	1	0	0	1	8	0	3	3	4	0	2
2	0	0	0	1	1	1	5	0	1	0	2	0	3
1	0	0	3	1	5	4	12	3	3	4	3	12	18
	72	72	72	72	72	72	72	72	72	72	72	72	72

7, Complete independence; 6, modified independence; 5, supervision; 4, minimal contact assistance; 3, moderate assistance; 2, maximal assistance; 1, total assistance

No impairment was found in cognition, with maximal scores in communication and social cognition items. The FIM™ items of cognitive assessment are related to activities in daily living and suffer a ceiling effect. When comparing the two groups, significant differences were found in age at assessment. This could be due to either age being an additional factor in disability or an independent risk factor for a worse outcome. When comparing age at onset, group 1 was found to have a higher average age, but the difference was not statistically significant. It is important to point out that some authors associate the onset of disease at older ages to a higher progression of disability motor rates.[28,29] The well-known variability in the progression of the disease may in part explain the lack of significant differences between groups regarding the duration of the disease.[28,30,31] Thus, duration does not necessarily implies greater disability.

Low back pain is another variable with significant statistical difference between the groups. Low back pain is a prevalent symptom[23] and can be easily linked to lower level of activity and higher degree of disability.

Paraparesis is the main aspect of this disease. Therefore, better motor scores will enhance patient activity level and independence.

Unlike traumatic spinal cord injury, HAM/TSP rarely presents a marked sensory-motor level and significant sensory deficits. Therefore the defining items that determine function are the motor scores.

Conclusions

Age, strength in the lower limbs and low back pain interfere with functional activities in patients with HAM/TSP. Duration of the disease is not a significant factor in patient disabilities. These patients showed no impairment in FIM™ cognition items and showed a high independence rate in some FIM™ items such as eating, grooming, bathing, dressing, toileting, bowel management and mobility; lower scores were found in locomotion (walk and stairs) and lowest scores prevailed in bladder management. The goals of rehabilitation treatment should target modifiable factors, such as pain, strength and neurogenic bladder management. The training of ADLs should be individualized, regarding level of disability, with priority to training independence in bladder management, transfers, gait and stairs. Further studies should be undertaken in order to better assess the results of this focused approach.

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