

Rehabilitation in Inclusion Body Myositis associated with human immunodeficiency virus (HIV) infection

Reabilitação na Miosite por Corpos de Inclusão associada a infecção pelo vírus da imunodeficiência humana (HIV)

Marco Orsini¹, Marcos RG de Freitas², Mariana Pimentel Mello³, Osvaldo JM Nascimento⁴, Eduardo Paranhos⁵, Carlos Henrique Melo Reis⁶

SUMMARY

Objective. To investigate the effects of a functional exercise program based on Proprioceptive Neuromuscular Facilitation techniques (PNF) on muscle strength and functional activities in a patient with inclusion body myositis (IBM) associated with human immunodeficiency virus (HIV) infection. **Method.** A patient with IBM was tested for muscle strength and functional capacities before and after a 16-week, patient-specific, home-based exercise program involving mild, daily and functional exercises. **Results.** Although real benefits of muscular force have been achieved, functional independence level was not modified. We believe the physical rehabilitation program served for minimization of the complications generated by the muscular weakness and optimization of motor abilities. **Conclusion.** The findings of this study indicate that an individually prescribed home exercise program, based on PNF techniques, can be safely implemented, since it respects the particularities of the patients and the disease, and is therefore beneficial in the management of patients with IBM, especially associated with HIV infection.

Keywords: Inclusion Body Myositis. HIV. Rehabilitation.

Citation: Orsini M, De Freitas MRG, Mello MP, Nascimento OJM, Paranhos E, Reis CHM. Rehabilitation in Inclusion Body

The research was done in the Neuromuscular Disease Outpatient Division at Universidade Federal Fluminense - UFF. Department of Neurology, Antônio Pedro University Hospital and Deolindo Couto Neurologic Institute - UFRJ, Niterói, RJ, Brazil.

1. Undergraduate in Medicine, Associate Professor of Neurologic Rehabilitation and Scientific Initiation, Escola Superior de Ensino Helena Antipoff (ESEHA), Department of Neurology, Federal Fluminense University - UFF, Niterói, RJ, Brazil.
2. Neurologist, Head of Staff, Department of Neurology, UFF, Niterói, RJ, Brazil.
3. Physical Therapist, Program of Scientific Initiation, Neurology Department, UFF, Niterói, RJ, Brazil.
4. Neurologist, Chair of Neurology, Neurology Department, UFF, Niterói, RJ, Brazil.
5. Physical Therapist - Program of Scientific Initiation, Neurology Department, UFF, Niterói, RJ, Brazil.
6. Neurologist, Chair of Neurology, Nova Iguaçu University - UNIG, Nova Iguaçu, RJ, Brazil.

RESUMO

Objetivo. Investigar os efeitos de um programa de exercícios funcionais com base nas técnicas de Facilitação Neuromuscular Proprioceptiva (FNP) na melhora da força muscular e nas habilidades cotidianas em um paciente com miopatia por corpos de inclusão (MCI) associada à infecção pelo vírus da imunodeficiência humana (HIV). **Método.** A força muscular e as habilidades funcionais foram testadas antes e após 16 semanas de treinamento a base de exercícios funcionais domiciliares diários de intensidade leve. **Resultados.** Embora ganhos de força muscular tenham sido obtidos em determinados grupamentos musculares, o nível de independência funcional não se modificou. Acreditamos, entretanto, que programa fisioterapêutico serviu para minimização das complicações geradas pela fraqueza muscular e na otimização das habilidades motoras. **Conclusão.** Os achados do estudo indicam que um programa domiciliar individualizado baseado nas técnicas de FNP pode ser implementado com segurança, desde que respeite as particularidades dos pacientes e da própria doença, tendo um efeito benéfico em pacientes com MIC, em especial, na associada à infecção pelo HIV.

Unitermos: Miosite por Corpos de Inclusão. HIV. Reabilitação.

Citação: Orsini M, De Freitas MRG, Mello MP, Nascimento OJM, Paranhos E, Reis CHM. Reabilitação na Miosite por

Endereço para correspondência:
Marco Orsini
R. Prof. Miguel Couto, 322/1001
24230-240 Niterói, RJ
E-mail: orsini@predialnet.com.br

Recebido em: 21/11/07
Revisado em: 22/11/07 a 04/05/08
Aceito em: 05/05/08
Conflito de interesses: não

INTRODUCTION

Sporadic inclusion body myositis (sIBM), a common adult-onset myositis, is characterized by an antigen-driven inflammatory response and vacuolar degeneration. The cause is unknown¹. The course of the condition is insidious, with gradually progressive muscular weakness and atrophy, which is typically selective in nature. Most severely involved are the quadriceps femoris muscles in the lower limbs, with a resulting tendency to falls, and the forearm flexor and extensor muscles, resulting in progressive weakness of the hands and impairment of manual control²⁻⁵.

The role of exercise therapy in IBM has received relatively little attention. Exercise has been shown to improve muscle strength, endurance, and well-being in patients with polymyositis, dermatomyositis and others neuromuscular diseases^{6,7}. However, there has also been concern that inappropriate levels of exercise could increase the degree of muscle damage and enhance the inflammatory process^{8,9}. Because of the variability in the degree of weakness, level of endurance, and general level of fitness among patients with IBM, it is important that any exercise program should be designed for the individual, that the initial exercise load should not be excessive, and the exercise program should be incremental and take into account not only gains in strength but also the overall functional capacity of the patient⁶. We report the association of sIBM with human immunodeficiency virus (HIV) infection and proposed some physical therapy strategies based on proprioceptive neuromuscular facilitation (PNF) techniques for clinical and functional management. The main objective of physical therapy appears to be the preservation of optimal quality-of-life throughout the course of this process.

METHOD

Case Report

Man, 56 years, retired cooker, relates the beginning of a picture of muscular weakness in the first semester of 1995, when he received the confirmation of HIV seropositivity. The diagnostic of IBM was upon the characteristic pattern of muscle weakness and atrophy and was confirmed by muscle biopsy. With passing of the years new functional limitations had been emerging, mainly in the execution of the standards of walk, to go up and go

down the stairs and in the execution of functional activities related with elevation of the upper limbs.

Experimental Design

After initial evaluation carried out in August of 2007 it was evidenced a picture of muscle weakness (Table 1) associated to the deficiencies/incapacities in daily activities (Table 2). For evaluation of muscular force it was used the scale established by the Medical Research Council¹⁰. Thirteen muscles were selected for test (Biceps brachii; Extensor carpi radialis; Triceps brachii; Fingers flexors; Dorsal and Palmar interosseous; Iliopsoas; Quadriceps femoris; Tibialis anterior; Extensor hallucis longus; Ankle plantar flexors; Gluteus Maximus; Adductors e Abductors of Tight). The application of the Functional Independence Measure¹¹ served to evaluate the impact of IBM on the daily life activities. The instrument evaluated 18 categories scored of one to seven and classified as regards the level of dependence for the achievement of specific tasks. Adding the points of the dimensions of MIF has gotten a total score minimum of 18 and the maximum one of 126 points that characterizes the levels of dependence for the subscores. The patient was reevaluated after 16 weeks of daily rehabilitative treatment in domiciliary environment and the results compared.

Table 1. Compromise of muscular force in the lower limbs by Medical Research Council¹⁰ (0. No movement; 1. Palpable contraction, no visible movement; 2. Movement but only with gravity eliminated; 3. Movement against gravity; 4. Movement against resistance but weaker than normal; 5. Normal power).

Muscles	Pre-Treatment Left/Right	Pos-Treatment Left/Right
Biceps brachii	4 / 4	4 / 4
Extensor carpi radialis	4 / 4	4 / 4
Triceps brachii	4 / 4	4 / 4
Fingers flexors	4 / 4	4 / 4
Dorsal and Palmar Interosseous	4 / 4	4 / 4
Iliopsoas	3 / 3	4 / 4
Quadriceps femoris	3 / 3	4 / 4
Tibialis Anterior	0 / 0	0 / 0
Extensor Hallucis Longus	1 / 1	1 / 1
Ankle Plantar Flexors	2 / 2	3 / 3
Gluteus Maximus	3 / 3	4 / 4
Adductors of Tight	3 / 3	4 / 4

Exercise Training Program

The diagonals associated to specific techniques of PNF were utilized as part of the training program (Table 3). The combination and the choice of the diagonals of movement was determined after a detailed evaluation of the muscular force in the upper limbs, lower limbs and trunk beyond the observation of the patient ability in the execution of 10 functional movements requested by the therapist: 1) to comb the hair, 2) to rise from a chair, 3) to brush the teeth, 4) to walk for 30 meters in leveled surface, 5) to go up and to go down stairs, 6) to remove the shirt; 7) to place the shirt; 8) to open the door handle of a door; 9) to search an object behind and in the high; 10) to sit down in the chair. The 10 diagonals selected for application (Table 3) and the respective specific techniques were selected after an accurate kinetic-functional diagnosis. After the accomplishment of the diagonals (3 series with 10 repetitions/day) the patient was stimulated to carry out the functions straightly associated to the movement standards. The training had duration of 4 months with a total of 100 services. The daily average time of the program was between 40–50 minutes, with pauses for rest (interval of 2 minutes between the movements). The patient carried through stretching before and after the activities. No movement provoked pain neither discomfort. Electrotherapeutic resources were not used. An only physiotherapist had carried through the sessions. We define as exercises of mild intensity those easily tolerated by the patient and not provocative of muscular weakness and fatigue. The approach of indirect treatment also was carried out, objectifying the irradiation of the muscular force of more fortified groupings for weakness muscles, unable to overcome the gravity and/or resistance imposed for the therapist. The training of position changes, transfers and balance had also consisted in the program of therapeutic exercises.

RESULTS

The muscular force according to the Medical Research Council evaluated before and after the treatment based on PNF techniques is presented in Table 1. The score of the FIM is before and after the treatment is presented in Table 2.

DISCUSSION

Neurologic disorders are frequent complications of HIV type 1 infection, and include central nervous system infections, neoplasms, vascular complications, peripheral neuropathies, and myopathies¹². Early series emphasized central nervous system (CNS) diseases, with relative few reports of primary disorders of peripheral nerve and muscle^{13,14}. This may be partially explained by the fact that coexisting central nervous system dysfunction such as dementia, focal brain lesions, or myelopathy may mask neuromuscular disorders. In advanced AIDS, the presence of peripheral neuropathy or myopathy may be overshadowed by other systemic conditions.

IBM has a slow progression, affects both the proximal and the distal muscles, and results in significant weakness and atrophy; sometimes they can be asymmetric, resembling a lower motor neuron disease¹. In typical cases muscle weakness and wasting are most profound in knee extensors, hip flexors and long finger flexors². Patients often present with

Table 2. Functional Independence Measure by Keith¹¹.

FUNCTIONAL INDEPENDENCE MEASURE (FIM)		
	BEFORE	AFTER
Self-Care		
A. Eating	7	7
B. Grooming	5	5
C. Bathing	5	5
D. Dressing Upper Body	5	5
E. Dressing Lower Body	5	5
F. Toileting	4	4
Sphincter Control		
G. Bladder Management	7	7
H. Bowel Management	5	5
Transfers		
I. Bed, Chair, Wheelchair Transfer	5	5
J. Toilet Transfer	5	5
K. Tub and Shower Transfer	5	5
Locomotion		
L. Walking / Wheelchair Locomotion	5 <input checked="" type="checkbox"/> W <input type="checkbox"/> WC	5 <input checked="" type="checkbox"/> W <input type="checkbox"/> WC
M. Stairs	4	4
Communication		
N. Comprehension	7	7
O. Expression	7	7
Social Cognition		
P. Social Interaction	7	7
Q. Problem Solving	7	7
R. Memory	7	7
TOTAL SCORE	102	102
Levels	Independent	
No Helper	7 - Complete Independence (Timely, Safely) 6 - Modified Independence (Device)	
Levels	Modified Dependence	
Helper	5 - Supervision (Subject = 100%+) 4 - Minimal Assist (Subject = 75%+) 3 - Moderate Assist (Subject = 50%+)	
	Complete Dependence	
	2 - Maximal Assist (Subject = 25%+) 1 - Total Assist (Subject = less than 25%)	

falls or difficulty performing certain tasks, such as turning keys, owing to quadriceps and finger flexors muscle weakness¹. Most patients require an assistive device, such as a cane, walker or wheelchair, within several years of onset³. The muscles of swallowing are also affected in IBM, and dysphagia is encountered by about 50% of the patients, leading to choking episodes². Sensory function is usually normal; mildly diminished vibratory sensation at the ankles is sometimes observed, but this is presumed to be age-related, or attributable to comorbidity. The tendon reflexes, although preserved in the early stages, can diminish in later stages when the atrophy of major muscle groups becomes evident¹.

Some of these characteristics are similar to those found in our case. Slow progression, with accentuated motor compromise in the flexors of the thigh, complicating the patient in the achievement of the normal standards of gait and to go up and go down the stairs. Happily, until the present, the muscles responsible for swallowing are preserved. The deep reflexes, in its majority, are hypoactive. As in the majority of the cases, the studied patient makes use of an assistance equipment use (orthotics) for locomotion and security in unlevelled surfaces, main cause of falls. After accomplishment of the physiotherapeutic program pre-established and application for a 6 months period, with a total of 100 sessions, we extract some conclusions. 1) Real benefits of muscular force have been achieved (Table 1) and functional independence (Table 2) was remained

stable. In addition the patient related more security, less fatigue and greater tolerance to determined functional activities. 2) We believe that our approach of treatment, based on the PNF techniques, served to minimize the secondary complications of IBM and, consequently, contributed for improvement the quality of life of the patient. 3) Another factor that deserves prominence was that even having in mind the progressive character of the illness, the efforts offered by the health professionals influenced positively in the life satisfaction and, mainly, fight against the illness.

The basic facilitation procedures provide tools for the therapist to help the patient gain efficient motor function and increased motor control^{15,16}. The basic procedures can use to treat patients with any diagnosis and or condition, although a patient condition may rule out the use of some of them. Evidence based physiotherapy treatment is based upon external support of the therapeutic care/ intervention combined with the expertise and experience of the therapist and adapted to the needs and objectives of the patient. Several articles can be used for supporting the choice of PNF treatment when the proper patient and treatment situation is present¹⁷⁻²². The results presented reinforce that the techniques of PNF, when employed after a correct kinetic-functional diagnosis, promote satisfactory results in the management of the muscular weakness and training of the functional abilities, in a patient with IBM associated to the infection by HIV. Despite of the physiotherapeutic intervention not

Table 3. Physical rehabilitation program established by the authors.

Rehabilitative Program – (PNF) (Diagonals of Movement)	Specific Techniques	Training Frequency	Series and Repetitions Pre-established	Associated function
Flexion-Adduction – External Rotation	Rhythmic Initiation	Daily	3 series/10 repetitions	To comb the hair
Extension-Adduction – Internal Rotation with Elbow Flexion	Rhythmic Initiation	Daily	3 series/10 repetitions	To wear the shirt
Flexion-Abduction – External Rotation	Rhythmic Initiation	Daily	3 series/10 repetitions	To remove the shirt
Flexion-Adduction – External Rotation with Knee Flexion and Trunk Patterns	Combination of Isotomics	Daily	3 series/10 repetitions	To rise from a chair
Diagonals of Lower limbs, MMII, Pelvis and Trunk.	Rhythmic Initiation	Daily	3 series/10 repetitions	Walk (30 meters)
Flexion-Adduction – Internal Rotation	Rhythmic Initiation	Daily	3 series/10 repetitions	To brush the teeth
Extention-Adduction – Internal Rotation	Rhythmic Initiation	Daily	3 series/10 repetitions	To open the door handle
Flexion-Abduction – External Rotation Flexion-Adduction – Internal Rotation	Rhythmic Initiation	Daily	3 series/10 repetitions	To search an object in the high
Diagonals of Lower limbs, MMII, Pelvis and Trunk	Combination of Isotomics	Daily	3 series/10 repetitions	To sit down in the chair
Diagonals of Lower limbs, MMII, Pelvis and Trunk	Rhythmic Initiation	Daily	3 series/10 repetitions	To go up and to go down stairs

to provoke improvement on the functional independence, a mild improvement in the clinical condition of the patient had been reached. The program acted in the control of the physical deconditioning and the muscle atrophy by disuse⁸. It is important stand out that the physiotherapist must carefully monitor the program of exercises or activities of the patient, to assure that any reduction in the force is more related with the progression of the illness than with the overuse of weakened muscles^{8,9}.

CONCLUSION

In respect to the nowadays view of evidence based treatment of neurological conditions, scientific support of our actions as a physiotherapist is necessary. There is a small amount of support for the PNF concept as an approach for physical rehabilitation. It becomes necessary new studies with a longer time of intervention and more expressive casuistic.

REFERENCES

1. Dalakas MC. Sporadic inclusion body myositis - diagnosis, pathogenesis and therapeutic strategies. *Nat Clin Pract Neurol* 2006;2(8):437-47.
2. Oldfors A, Lindberg C. Diagnosis, pathogenesis and treatment of inclusion body myositis. *Curr Opin Neurol* 2005;18:497-503.
3. Peng A, Koffman BM, Malley JD, Dalakas MC. Disease progression in sporadic inclusion body myositis: observations in 78 patients. *Neurology* 2000;55:296-8.
4. Dalakas MC. Inflammatory, immune and viral aspects of inclusion-body myositis. *Neurology* 2006;66(Suppl):S33-8.
5. Cupler EJ, Leon-Monzon M, Miller J, Semino-Mora C, Anderson TL, Dalakas MC. Inclusion body myositis in HIV-1 and HTLV-1 infected patients. *Brain* 1996;119:1887-93.
6. Alexanderson H, Lundberg IE. The role of exercise in the rehabilitation of idiopathic inflammatory myopathies. *Curr Opin Rheumatol* 2005;17:164-71.
7. Wiesinger GF, Quittan M, Nuhr M, Volc-Platzer B, Ebenbichler G, Zehetgruber M, et al. Aerobic capacity in adult dermatomyositis/polymyositis patients and healthy controls. *Arch Phys Med Rehabil* 2000;81:1-5.
8. Alexanderson H, Stenström CH, Lundberg I. Safety of a home exercise programme in patients with polymyositis and dermatomyositis: a pilot study. *Rheumatol* 1999;38:608-11.
9. Heikkilä S, Viitanen JV, Kautianen H, Mänttyvuo P, Harju T. Rehabilitation in myositis. *Physiother* 2001;87:301-9.
10. Medical Research Council. Aids to the investigation of peripheral nerve injuries. War Memorandum. 2nd edition. London: HMSO, 1943, 1-2.
11. Keith RA, Granger CV, Hamilton BB, Sherwin FS. The functional independence measure: a new tool for rehabilitation. *Adv Clin Rehabil* 1987;1:6-18.
12. Simpson DM, Tagliati M. Neurologic Manifestations of HIV Infection. *Ann Int Med* 1994;121(10):769-85.
13. Snider WD, Simpson DM, Nielsen S, Gold JW, Metroka CE, Posner JB. Neurological complications of acquired immunodeficiency syndrome: Analysis of 50 patients. *Ann Neurol* 1983;14:403-18.
14. Levy RM, Bredesen DE, Rosenblum ML. Neurological manifestations of the acquired immunodeficiency syndrome (AIDS): Experience at UCSF and review of the literature. *J Neurosurg* 1985;62:475-95.
15. Ferber R, Gravelle DC, Osternig LR. Effect of PNF stretch techniques on trained and untrained older adults. *J Aging Phys Act* 2002;10:132-42.
16. Adler S, Beckers D, Buck M. PNF in Practice. 2nd ed. Berlin: Springer, 2003, 401.
17. Johnson GS, Johnson VS. The application of the principles and procedures of PNF for the care of lumbar spinal instabilities. *J Manual Manipul Ther* 2002;2:83-105.
18. Klein DA, Stone WJ, Phillips W, Gangi J, Hartman S. PNF training and physical function in assisted living older adults. *J Aging Phys Act* 2002;10:476-88.
19. Wang RY. The effect of proprioceptive neuromuscular facilitation in case of patients with hemiplegia of long and short duration. *Phys Ther* 1994;12:25-32.
20. Kofotolis N, Eleftherios K. Effects of two 4-week PNF programs on muscle endurance, flexibility, and functional performance in women with CLBP. *Phys Ther* 2006;7:1001-12.
21. Kofotolis N, Kellis E. Effects of two 4-week proprioceptive neuromuscular facilitation programs on muscle endurance, flexibility, and functional performance in women with chronic low back pain. *Phys Ther* 2006;86(7):1001-12.
22. Schuback B, Hooper J, Salisburg L. A comparison of a self stretch incorporating PNF components and a therapist applied PNF technique on hamstring flexibility. *Physiother* 2004;3:151-7.