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The Effectiveness of an Individualized, Home-Based Functional Exercise Program for Patients With Sporadic Inclusion Body Myositis

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Abstract

Objective:
The objective of the study was to investigate the effects of a functional exercise program on muscle strength and mobility in a group of patients with sporadic inclusion body myositis (IBM).

Methods and Materials:
Seven patients with sporadic IBM were tested for muscle strength and functional capacities before and after a 16-week, patient-specific, home-based exercise program involving mild, daily, functional exercises.

Results:
Significant improvements in isometric strength were demonstrated in all muscle groups tested and were maximal in the hip flexor muscles. In addition, walking and stair climbing times improved in all patients. The exercise program was well-tolerated, and there was no significant change in the serum creatine kinase level following the exercise period.

Conclusion:
The findings of this study indicate that an individually prescribed home exercise program can be safely implemented and can result not only in significant gains in muscle strength but also in useful improvements in functional capabilities and is therefore beneficial in the management of patients with IBM.

Key Words: inclusion body myositis, exercise program, muscle strength, functional capabilities

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INTRODUCTION

Sporadic inclusion body myositis (IBM) is the most common form of inflammatory myopathy in people older than age 50 years, but it also can affect younger individuals.[1–3] 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 control.[4–8] The condition is usually unresponsive to treatment with corticosteroids and immunosuppressive agents and most patients need an assistive walking device after 5–10 years or become confined to a wheelchair.[9–11] As the disease progresses and the degree of mobility diminishes, additional factors such as muscular and cardiovascular deconditioning and disuse atrophy of muscles contribute to the overall level of disability.

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 and dermatomyositis.[4,12,13] However, there has also been concern that inappropriate levels of exercise could increase the degree of muscle damage and enhance the inflammatory process.[4,14] Two previous studies of resistance exercise training in small groups of patients with IBM failed to show any significant improvement in isometric muscle strength,[15,16] although in 1 study dynamic strength and maximal torque

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production improved, particularly in clinically less affected muscles,[15] without changes in the serum creatine kinase (CK) level or an increase in histological markers of disease activity.

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 that the exercise program should be incremental and take into account not only gains in strength but also the overall functional capacity of the patient. The purpose of the present study was to investigate the effects of such a patient-specific, home-based, incremental exercise program on muscle strength, endurance, and functional capabilities in a group of patients with sporadic IBM.

MATERIALS AND METHODS

Patients

Seven patients (4 male, 3 female; mean age 67.3 years; disease duration 4–17 years; Table 1) with sporadic IBM were recruited for the 16-week exercise program from the Neuromuscular Clinic at the Australian Neuromuscular Research Institute (ANRI) in Perth, Western Australia. The diagnosis was based upon the characteristic pattern of muscle weakness and atrophy and was confirmed in all cases by muscle biopsy. All had been in a state of progressive decline in muscle strength and function and overall functional ability, and all conformed to the diagnostic criteria for definite IBM.[1] The study was approved by the Ethics Committee of the Sir Charles Gardiner Hospital, and patients signed an informed consent prior to beginning the exercise program.

Experimental Design

Muscle strength and functional assessments were performed and serum CK levels were measured before and after completion of a 16-week training period. A Penny and Giles handheld myometer (Penny and Giles Transducers, Christchurch, England), a valid and reliable instrument for muscle-strength testing in patients with IBM,[13] was used to measure muscle strength using the "make" method in the following muscles bilaterally: shoulder abductors and external rotators; elbow flexors and extensors; wrist extensors; hip flexors and abductors; and knee flexors and extensors. Grip strength was measured using a Speidel and Keller, Stabil 3 grip dynamometer (Speidel and Keller, Jungingen, Germany). Medical Research Council (MRC) ratings were also recorded preintervention and postintervention using a 0–5 rating scale.[17]

The following functional tasks were also evaluated: time taken to walk 30 meters and to walk up 1 flight of stairs and the maximum

TABLE 1. Patient Characteristics and Serum Creatine Kinase Levels Preexercise and Postexercise Intervention

Age	Knee Ext. MRC Preintervention	Ambulatory Assistance	Age at Disease Onset	Serum CK During the Exercise Program (U/L)	
				Start	End
66	3+	No	62	596	874
61	5	No	56	776	396
73	4+	Yes—motorized buggy	62	295	392
66	2	Yes—cane	61	198	147
68	3–4	Yes—motorized buggy	51	N/A	N/A
67	2–3	Yes—cane	56	223	225
73	3	No	58	342	220

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number of sit-to-stands performed by the patient in each exercise session during the training period. All assessments were performed by the same examiner (LGJ). Standard instructions and levels of positive reinforcement were provided to all patients.

Exercise Training Program

The range of exercises used is shown in Table 2. The combination of exercises prescribed for each patient was determined after a detailed assessment of limb and axial muscle strength and after observing the patient's ability to perform a variety of functional movements. Multijoint exercises such as moving from the sitting to the standing position were employed in all patients. Also, varying combinations of the other exercises were included, selected on the basis of the degree of weakness of proximal muscle groups and functional impairment reported by the patient and observed by the examiner. The sitting-to-standing exercise was selected and prescribed to all patients because it involves the coordinated use of multiple muscle groups to rise from the seated position, including the elbow extensors, latissimus dorsi, and quadriceps femoris, with forward momentum generated by the rectus abdominis and hip flexors. Additionally, certain patients undertook isometric vastus medialis exercises to further train the quadriceps muscle group. The exercises were initially demonstrated to the patient by the investigator, who then supervised the patient performing the exercises to ensure that the correct technique was being used.

The exercise load during the first 2 weeks of the program was kept at a low level to prevent overloading. Patients were instructed to perform the set of exercises twice daily on each day of the week but were told to abstain from exercising on any given day if they were still experiencing residual fatigue or muscle soreness from the previous day's exercises. The number of repetitions of each exercise was 2 less than the maximum number of times that they were able to repeat the exercise in the initial assessment session. Patients reported back by telephone at the end of the initial 2-week period. Depending on the patient's tolerance to the initial exercise load, they were instructed to progressively increase the exercise load first by increasing the number of exercises (up to a maximum of 6); followed by the number of repetitions of each exercise (up to a maximum of 15); and, last, by the number of sets of each exercise (up to a maximum of 3). Only 1 of the variables (namely, number of exercises, number of repetitions, or number of sets) was increased in any 1 fortnight.

TABLE 2. Range of Exercises Prescribed and the Individualized Exercise Program Progression of Subject 5

Exercises	Preintervention	Postintervention
1. Whole body		
Sitting to standing		
(from standard height chair with arms)	3 sets of 6/day	3 sets of 10/day
2. Upper limbs		
Biceps curls*	2 sets of 10/arm/day	2 sets of 10/arm/day
Shoulder presses*		
Seated rowing (Thera-Band)		
Wrist flexion/extension*		
3. Lower limbs		
Calf raises (on tiptoe)		1 minute 2/day
Calf stretches (against wall)		15–20 seconds 3/day
Vastus medialis (isometric)		
Ankle dorsiflexion		2 sets of 20/day

*Holding a 375-g can of food in each hand.

Patient Monitoring

A description of the exercises to be undertaken and a data recording sheet were given to the patient to take home. Patients were instructed to fill out the sheet each day, recording the exercises performed and their respective volumes and frequency. They were also asked to assign a value to the degree of fatigue, soreness, and breathlessness (based on a 1–10 scale) experienced during the exercises and following the completion of each day's tasks.

Each patient was counseled on the importance of doing the exercises as shown; of undertaking the exercises to the volume, intensity, and frequency prescribed; and of recovering from the exercises and not undertaking other irregular strenuous activities. Progress during the exercise period was monitored by regular phone calls and scheduled visits to the Neuromuscular Clinic. At the completion of the 16-week training period, the patients attended the Neuromuscular Clinic for muscle strength and functional assessments and serum CK estimation.

Data Analysis

Raw data for muscle strength tests, stair and walk time, and number of paces taken to complete the walk were assessed for normality (Kolmogorov-Smirnov) across subjects. Pre intervention and postintervention comparisons for each variable were performed using a paired *t*-test (1 tailed) for normally distributed data and a Wilcoxon matched-pairs signed-ranks test for data not normally distributed. The level of significance was set at $P < 0.05$. Postintervention data were normalized to preintervention levels for each subject, before being averaged across the group. Group data are reported as mean ± standard error of the mean (SEM).

RESULTS

Strength Assessments

A significant improvement was seen in mean muscle strength in both the upper and lower limbs after the functional exercise program. Figure 1 shows that all muscles tested increased in strength following the intervention period, ranging from 171 ± 56% in the hip flexors to 23 ± 8% for the shoulder external rotators. Differential improvements occurred at the knee (flexors more than extensors), hip (flexors more than abductors), shoulder (abductors more than external rotators), and elbow (extensors more than flexors). Grip strength also significantly improved by an average of 52 ± 32% following the exercise intervention. MRC grades in individual patients remained unchanged at the end of the 16-week exercise period. Table 3 shows group-averaged muscle strength data preexercise and postexercise intervention, with improvements in all muscle groups following the 16-week training intervention.

Functional Assessments

The time taken to climb a flight of stairs and to walk unaided for 30 meters improved

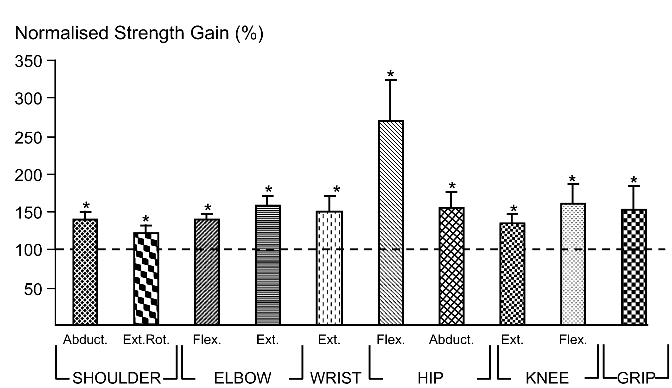

FIGURE 1. Mean percentage change in patients (n = 7) muscle strength following the exercise intervention when compared to pre-intervention strength levels normalized to 100%. *$P > 0.05$

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TABLE 3. Mean and Standard Error of Group (n = 7) Muscle Strength (kg-f) Preexercise and Postexercise Intervention

	Right		Left	
	Pre	**Post**	**Pre**	**Post**
Shoulder abduction	8.7	12.7	8.8	11.8
	1.1	1.9	0.8	1.7
Shoulder external rotation	6.2	7.4	5.9	7.0
	0.8	0.6	0.7	0.7
Elbow extension	6.2	8.2	5.1	7.1
	0.8	1.1	0.7	0.9
Elbow flexion	7.8	10.6	5.5	10.1
	1.2	1.9	0.7	1.5
Wrist extension	7.9	10.9	6.5	9.6
	0.9	1.2	0.6	1.3
Hip flexion	6.8	13.9	5.7	13.8
	1.0	1.4	0.8	1.8
Hip abduction	6.6	9.6	5.7	8.1
	0.9	1.1	0.9	0.9
Grip strength	156	162	116	144
	38	36	41	42
Knee extension	6.4	8.5	5.8	8.9
	1.1	1.6	0.8	2.0
Knee flexion	6.7	10.4	6.1	9.4
	0.9	1.6	0.9	1.4

in 6 patients and was unchanged in 1 (group average 14 ± 3 versus 11 ± 3 seconds; preintervention versus postintervention) (Fig. 2). The time taken to complete the 30-meter walk improved in all patients and was reduced by 17% in the group as a whole (41 ± 5 versus 34 ± 5 seconds; preintervention versus postintervention). There was also a small but nonsignificant reduction in the number of paces performed during the 30-meter walk following the training period (from 59 ± 3 to 55 ± 4). Four of the 7 patients improved in their capacity to perform sit-to-stands, 2 maintained their preintervention capacity, whereas 1 experienced a slight decrement (Fig. 3).

Subjective Changes

All 7 patients adhered to and completed the prescribed exercise program, with compliance to the prescribed exercise training plan by the patient group estimated at 90%–95%. Only 2 reported minor muscle soreness and fatigue during the exercise sessions, but none reported persisting symptoms. There was no formal assessment of changes in quality of life or capacity to perform activities of daily living, but patients were questioned on their subjective feelings of changes to their lives as a result of the exercise intervention during the posttesting appointment. Each patient felt that they had derived a benefit from the exercise training in terms of general health and well-being.

Serum CK Levels

There were only minor changes in CK level after completion of the exercise program (Table 1). The mean CK level in the group was 405 ± 94 U/L pre-exercise and 375 ± 108 U/L post-exercise.

DISCUSSION

It has previously been thought that exercise programs should be avoided in patients with inflammatory myopathies because of concern that the exercise could aggravate the underlying inflammatory process.[4,14] However, studies in other forms of

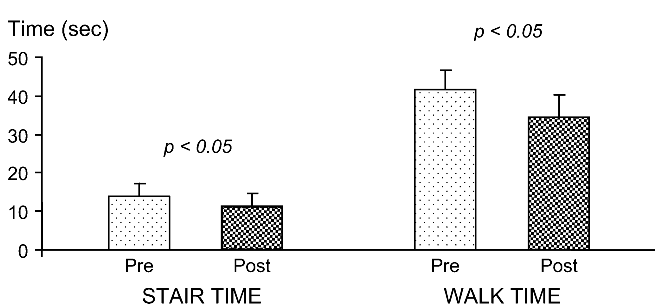

FIGURE 2. Mean changes in time taken to climb one flight of stairs and walk unaided for 30 meters following a 16-week exercise training intervention (n = 7).

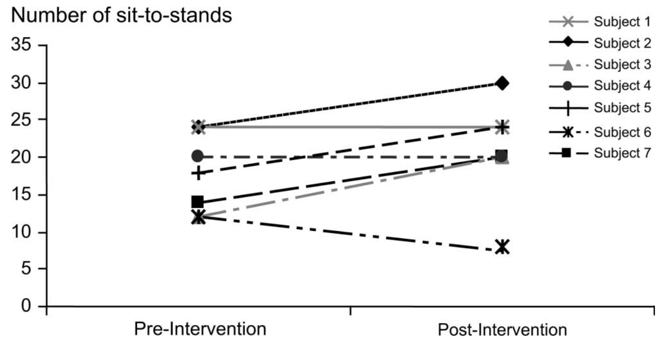

FIGURE 3. Individual changes in number of completed sit-to-stands per day over the 16-week intervention period.

idiopathic inflammatory myopathies, such as polymyositis and dermatomyositis, showed a positive response to physical training in the absence of an adverse effect on the disease process.[12,18] Furthermore, studies with patients with IBM using strength and aerobic training concluded that exercise can be performed safely, can lead to dynamic strength improvements, and possibly can prevent continued loss of muscle strength.[15,16] However, optimal exercise programs resulting in improvements in muscle strength and function have not been established in either a home-based or clinic setting. The emphasis of this study on selecting a set of exercises designed to improve limb and whole-body function, including the use of multijoint exercises and the integration of balance and stabilizing components, distinguishes this study from previous exercise intervention studies in IBM.

This study has shown that a closely monitored, 16-week, home-based, individualized functional exercise program can lead to significant gains in muscle strength and improvements in the performance of functional tasks in patients with IBM. The patient-specific aspect of this study, and the prescription of exercise on a low-intensity, high-frequency basis, allowed the investigator to gently overload each patient while targeting the muscles primarily affected by the disease.[19] The protocol was well tolerated by all the patients and did not cause adverse muscle symptoms or elevation of serum CK levels. From interviews with the patients, it was generally felt by those who had previously had exercise therapy that this had been too strenuous and had resulted in muscle pain and increased levels of weakness and fatigue. A previous study on the impact of a home–based physical activity in people with neuromuscular diseases also indicated that a conservative approach, as used in the present study, is associated with better program adherence and reduces excessive muscle fatigue and soreness, which were identified as major determinants for withdrawal.[20]

Class-based versus home-based exercise rehabilitation therapy has been investigated in a variety of pathological groups,[21] and the cost-effectiveness, compliance rates, and overall results from both strategies have been found to have merit.[22] Home-based programs are easy to implement, are accessible to the patient, and encourage compliance.[19] The impaired mobility that typifies a patient with IBM gives a home-based exercise program obvious advantages over a class-based program—in particular, the safety and familiarity of exercising in their own home, the assistance and encouragement of partners, the flexibility to perform the exercises according to their own schedule, and the avoidance of arranging transport.

Because postexercise muscle biopsies were not performed we do not know whether the improvement in isometric strength was associated with changes in muscle fiber sizes or in the proportions of different fiber types. However, in a previous 12-week trial of resistance exercise training in IBM an increased

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cross-sectional area of Type I muscle fibers and an increased percentage of Type IIc fibers was found in postexercise biopsies.[16] The improved muscle strength could therefore be the result of hypertrophy of Type I fibers, or of fibers that had undergone atrophy as a result of disuse or of the disease process. Other possible explanations for the gains in functional performance include motor learning and improved coordination of muscles activated during the performance of a task. We hypothesize that the present approach has beneficial effects by slowing or reversing muscle fiber atrophy and by improving coordination and activation of muscle groups to perform a given task. The use of functional weightbearing activities in the exercise programs may help to induce performance improvements in everyday living activities.[23] It is also hypothesized that strength gains in muscle groups not specifically trained in isolation, such as the hip flexors, was a result of their secondary involvement in the prescribed exercises, such as the sit-to-stand exercise. The results are particularly promising when taking into consideration that the training did not include an aerobic component that could further enhance the patient's ability to move with a greater efficiency.

In conclusion, we have shown that an individualized, supervised program of mild daily exercises performed in the patient's own home can lead to significant increases in muscle strength and to useful functional improvements in walking and mobility in patients with IBM, even in those with longstanding disease. Such changes have positive implications for the patients' overall well-being and capacity to perform activities of daily living. Refinement of training protocols and the implications they have on the patient's quality of life requires further investigation using larger sample sizes in controlled, long-term training interventions.

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