## **CASE STUDY**



# OUTPATIENT PHYSICAL THERAPY EVALUATION AND TREATMENT OF A PATIENT DIAGNOSED WITH SPORADIC Inclusion Body Myositis: A case study

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# ABSTRACT

**Background:** Sporadic inclusion body myositis is an autoimmune and degenerative disorder of skeletal muscle that affects people at random. It most commonly begins as progressive weakness and atrophy of lower extremity musculature, beginning with the proximal leg. These impairments in body structure adversely affect the performance of functional activities and mobility, resulting in a progressive decrease in independence and participation both at home and in the community. Physical therapy attempts to minimize these effects through educational and procedural interventions focused on treating impairments and limitations. The purpose of this case study was to provide a description of the physical therapy management of a patient diagnosed with sporadic inclusion body myositis.

*Case Summary:* The patient was a 66-year-old male who was diagnosed with sporadic inclusion body myositis with a chief complaint of weakness and fall risk. He presented with generalized lower extremity weakness and atrophy of bilateral quadriceps, as well as impaired balance and increasing fatigue with activity. Therapeutic exercise, home exercise program, balance, gait, and stair training were delivered to address these impairments. Patient outcomes showed improvement in balance and safety with functional activities.

*Discussion:* The patient was seen for seven visits that were 45 – 60 minutes in length, over a five-week period. The patient made subjective reports of improvement in functional activities and balance; however many objective outcome measures could not be reassessed. There is a need for further research on this population to determine the effectiveness and parameters of physical therapy interventions.

*Conclusion:* Physical therapy may have helped improve balance as well as subjective reports from the patient of increased feeling of confidence while navigating stairs.

*Keywords:* Myositis, Physical Therapy, Balance, Rehabilitation, Weakness.

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## **INTRODUCTION**

Sporadic inclusion body myositis is an autoimmune disease and degenerative disorder of the skeletal muscle [1,2]. It is the most frequently acquired myopathy in patients over 50 years of age [2-4]. There are two types of inclusion body myositis: Inflammatory inclusion body myositis and Hereditary inclusion body myositis. The inflammatory version is divided into two types. Sporadic affects people at random, and there is idiopathic inflammation of the skeletal muscle. The second type is familiar, and it is characterized because it affects multiple siblings but is not passed on through the generations.

Prevalence of sporadic inclusion body myositis is from 1-71 per million people. When limited to people over the age of 50 the prevalence increases to 139 per million. There are 5-11 per million people affected by it in the United States [4,5]. It is classified as a rare disease in the United States. It predominantly affects males with a ratio of 2:1 to 3:1 [7]. It rarely affects people under the age of 45. Patients report a 5 to 8-year delay from the initial presentation to the final diagnosis [7]. The etiology of inclusion body myositis is primarily unknown. There is evidence to support both the autoimmune and neurodegenerative theories.

Clinical manifestations of the condition show insidious onset with progressive muscular weakness in the proximal leg and the distal arm. It most commonly affects the lower extremity leading to increased falls [6-8]. Patients have difficulty standing and lose finger flexor strength and dexterity. Dysphagia also affects 50-70% of patients. The 2011 European Neuro Muscular Center (ENMC) diagnostic criteria include duration of weakness greater than 12 months, Creatine Kinase levels no greater than 15 times the upper normal limit, age at onset greater than 45 years, and finger flexor weakness greater than shoulder abduction weakness or knee extension weakness greater than hip flexion weakness. [9-11] High doses of Prednisone, Methotrexate, or other medications did not slow the process of the disease and resulted in increased impairments. [12-15]

The prognosis demonstrates that patients will require an assistive device approximately five years after the diagnosis and may require a wheelchair ten years after the diagnosis. Over half of the patients affected with the condition reported inability to ascend stairs.

Physical therapy may be needed to manage the progressive symptoms of myositis. Studies conducted have shown aerobic and strengthening exercises are safe in this population [16-19]. There is no increase in creatine kinase levels, no increased signs of inflammation, and no reduction in muscle strength. Physical therapy interventions may include: aerobic training, gait training, balance, stretching, strength training, energy conservation, and assistive device training. The success of specific interventions has not been studied in this population [6,17,18,19]. The purpose of the case study was to provide a description of the physical therapy management of a patient with sporadic inclusion body myositis.

## **Patient Information**

The patient was a 66-year old with sporadic inclusion body myositis. He had been retired four years previously from mail delivery. He worked part time driving a school bus. His primary complaint was a weakness of both lower extremities. He reported atrophy of his quadriceps bilaterally, difficulty getting out of chairs, inability to squat to the floor to reach objects, and increased fatigue. Even though the patient showed reduced ambulation velocity, weakness with ankle dorsiflexion, instability on stairs, frequent loss of balance, and inability to ambulate long distances, aerobic and strengthening exercises are safe in this population. [16-20]

The patient's history of the present illness started with the weakness of the lower extremities gradually over five years. He noticed atrophy of the quadriceps and difficulty squatting and moving from sitting to standing. He also demonstrated impaired balance with two falls on the stairs within the last six months. His participation in hunting, fishing, and farming activities diminished as well. The patient was referred from a physician to evaluate and treat for muscle weakness and reduced balance. The patient's past medical history included hernia repair. The patient was taking Glucosamine HCL and Omeprazole. His living environment consisted of a four level home with two steps without a railing to enter the house and two flights of stairs with a railing in the home. The patient was in the process of building a one level home due to the medical condition. He lived with his wife. He enjoyed fishing several times each month and hunting throughout the year. He worked on the farm each weekend during the summer completing chores like spraying weeks, lawn care, and maintenance requirements. He enjoyed playing with grandchildren every day. The patient wanted to be able to get into larger vehicles more easily, ride a bike 2 miles, get out of low chairs, and have no falls on the stairs.

#### **Review of Systems**

The patient reported occasional dizziness when standing associated with the cardiopulmonary system. The musculoskeletal system review was affected by weakness and reduced motion. There was reduced balance in the neurological system. All other systems were at functional levels. The patient was anxious and had difficulty understanding instructions.

## **Initial Clinical Impression**

The patient was a good candidate for physical therapy to reduce functional risks at home and the community. No other referrals were made to other health care professionals.

#### **Physical Exam**

The patient was 5'9" and 218 pounds with a body mass index of 32. His vital signs were normal. His cervical and lumbar spine motion were limited bilaterally with side bending and rotation. Hamstring tightness was noted in both lower extremities. Dermatomes in all extremities were intact. Clonus was negative in all extremities.

## **Tests and Measures**

Observationally, the patient ambulated with a slow cadence, shortened step length bilaterally, forward trunk lean, minimal ankle dorsiflexion, narrow base of support, and reduced eccentric quadriceps control bilaterally. He had poor posture with a forward head, rounded shoulders, and increased kyphosis and lordosis. During transfers, the patient uses both upper extremities supported on his knees to come to a standing position. He is unable to complete a functional squat. Manual muscle testing is limited in the lower trapezius and serratus anterior bilaterally at 4/5. In the lower extremities, the patient is rated at 3+/5 in the gluteus maximus, gluteus medius, quadriceps, anterior tibialis, and extensor halluces bilaterally. Palpation reveals reduced muscle mass without pain in the lower extremities. The initial Berg Balance outcome measure was 55/56 with a minimal detectable change of 3.3 [21,22]. This indicates there is some loss of functional balance but not a risk for falls. The Single Leg Stance [23] with eyes open and no upper extremity support was 8 seconds on the right and 5 seconds on the left. The patient had a positive Trendelenburg sign bilaterally. Normative data suggests males of the same age as the patient perform the Single Leg Stance Test for 28.7 seconds on each lower extremity. The Lower Extremity Functional Scale (LEFS) was initially scored at 67/80 with a Minimal Clinically Important Difference of 9 scale points [24]. The Timed Up and Go Test was 7.5 seconds which was within the normative data for community-dwelling adults [25]. The Timed Up and Go test was 7.8 seconds. [26,27] For the Five Times Sit to Stand Test, the patient was unable to complete it without using his upper extremities. Using a modified technique using one hand on the knee and one hand on the table the patient could complete the task in 24.5 seconds [28]. Patients of the same age completed the task in 8 seconds.

## **Diagnostic and Assessment**

By the International Classification Framework (ICF) the patient's health condition was Sporadic Inclusion Body Myositis. The impairments were generalized lower extremity weakness, excessive fatigue, and reduced balance. The activity limitations were the inability to walk one mile, difficulty on stairs, inability to perform a full squat, and limited transfers. Difficulty participating in recreational activities, farm duties, and community ambulation were the participation restrictions. Environmental Factors were: a supportive wife, building a new single level home, frequently works on family farm, rides a bicycle to part time job of a driving school bus. Personal Factors were: motivation to increase current functional levels, limited knowledge of the medical condition, recreational activities require walking long distances over rough terrain, overexertion at the family farm, adherence to a home exercise program, difficulty performing exercises correctly at home and in the clinic without significant levels of curing. Based on the evaluation information the patient was diagnosed with impaired strength, limited mobility, and fall risk associated with sporadic inclusion body myositis [6-8]. Physical therapy was indicated to improve function, strength, aerobic conditioning, patient education, and home exercise program [16-20]. The prognosis for the disease was limited since there was no known restorative treatment to slow the disease progression [1,2,6,7,8]. Physical therapy will minimize the effects of the condition by strengthening the unaffected musculature and improving aerobic conditioning based on the established goals. The plan of care addressed the functional impairments thus working towards independence with a home exercise program. The use of Rate of Perceived Exertion was used to limit muscle soreness and any eccentric loading of the lower extremities. The patient goals were as follows: Patient will improve eyes open single leg stance time to 30 seconds bilaterally to demonstrate improved balance (four weeks), 2) Patient will improve lower extremity manual muscle tests score 1/2 grade to 4/5 to demonstrate improved strength (six weeks), 3) Pt will report a clinically significant improvement of 10 points on LEFS score (eight weeks), 4) Patient will demonstrate independence with home exercise program (sixteen weeks), 5) The patient will report no falls with ambulation over ground or navigating stairs (sixteen weeks). The patient was instructed to continue riding his bike 2 miles to work over three days each week. The frequency of the visits was to be two times each week for 16 weeks for approximately 1 hour. Patient education, strengthening, balance training, gait training, stretching, and home exercise program were some of the major interventions used.

## Interventions

Patient Education was the primary intervention describing the condition, its progressive nature, avoiding strenuous activities, and the impact of exercise on the patient's overall condition and quality of life [6,7,8,16,20]. The home exercise program was a primary part of patient education. Strengthening involved low to moderate level exercises to avoid further injury to the affected muscles [20]. The patient was monitored for muscle fatigue and soreness with RPE below 13. No more than two sets of 10 repetitions was the initial exercise prescription [16,18,19]. NuStep, body weight exercises, resistance band exercises, ankle weight exercises, and core strengthening exercises were also incorporated. Balance training consisted of single leg stance training with eyes open on a solid surface and no upper extremity support with standby assist [6]. Three trials of 60 seconds were performed bilaterally with cues to prevent lower extremities from making contact and to maintain hands on the waist. No studies have been conducted within this population to determine the effectiveness of specific balance exercises [19]. Cone Reaches were performed bilaterally. The patient stepped forward as far as possible toward a cone target to improve longer step length and maximize single leg balance while stepping. After taking a step, the patient completed a partial lunge to challenge balance and strengthen the quadriceps. Gait training was completed over level surfaces for 8-10 minutes with comfortable walking speed, and standby assists for the physical therapist. Obstacles were also used with gait on level surfaces with standby assist for 5 minutes to step over thus simulating gait on rough terrain on the farm. The patient performed 12 stair training using a step through the pattern and one handrail to improve ascending and descending stairs for two bouts with standby assist and moderate cues for safety and eccentric control. Stretching was performed to address hamstring tightness and lower extremity mobility two repetitions holding the position for 60 seconds. The home exercise program included aerobic conditioning exercises, strengthening, and stretching at home [16,18,19,20]. The patient was instructed to perform the home program five times each week and one time each day on the days he did not have physical therapy. The patient was not to do the home program if he was experiencing muscle soreness or fatigue from the previous day activities. Gradual progression of interventions was the focus of the plan of care.

## Follow Up and Outcomes

The patient was treated seven times over five weeks. He reported increased confidence and feelings of stability while ambulating and using the stairs. Single leg stance improved to 50 seconds on the right lower extremity and to 30 seconds on the left lower extremity by the end of 5 weeks. He also made improvements in the number of repetitions in the sit to stand the test. The patient met the single leg stance goal and reported no falls over the five weeks of physical therapy. The patient made improvements toward achievement of the home exercise program goal requiring less assistance and cues but did not consistently perform the program between visits.

## DISCUSSION

The progressive nature of the patient's condition presents a unique challenge to physical therapists. Machado in 2014 and Johnson in 2009 are some of the studies that have been conducted looking at the efficacy of physical therapy interventions [6,16,17,18]. One of the facts that were found was there was some delay in the physical therapy visits due to health insurance issues. Delayed physical therapy may have affected the patients progress in the program. The implications of delayed rehabilitation may reduce the potential positive outcomes of patients with this condition. Another fact was that balance a positive intervention that improved the patient's confidence and functional activities at home. Balance may be a useful intervention for patients with sporadic inclusion body myositis. The implications for balance training may need additional study to determine greater generalization to this population. Another fact was the lack of patient education. This implies that future activities with more detailed patient education from the collaborative efforts of a multidisciplinary team may be of benefit. Machado et al. in 2014 suggest collaboration is critical to improving treatment for these patients [6]. Also, exercise prescription should be instituted more slowly to facilitate patient adherence. It would also be helpful to us the Inclusion Body Myositis Functional Rating Scale to assess progress toward goals and more independent function [2]. Future research should compare various physical therapy interventions to determine the efficacy of each and consider exercise progression rate.

## CONCLUSION

Physical therapy intervention may have been beneficial for a patient with Inclusion Body Myositis. The patient made improvements in confidence and reduced fall risk while participating in 5 weeks of physical therapy.

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|                                      | Week 2<br>Visit 2 | Week 2<br>Visit 3 | Week 3<br>Visit 4     | Week 4<br>Visit 5 | Week 4<br>Visit 6     | Week 5<br>Visit 7 |
|--------------------------------------|-------------------|-------------------|-----------------------|-------------------|-----------------------|-------------------|
| Nustep                               | Yes               | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| 3 position<br>straight leg<br>raise  | Yes               | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| Side lying abduction                 | Yes               | Yes               | D/C to<br>HEP<br>only |                   |                       |                   |
| Clamshells                           |                   |                   |                       |                   | Yes                   | Yes               |
| Seated<br>knee<br>flexion            | Yes               | Yes               | Yes                   | Yes               | D/C to<br>HEP<br>only |                   |
| Seated<br>ankle dor-<br>siflexion    | Yes               | Yes               | Yes                   | Yes               | D/C to<br>HEP<br>only |                   |
| Seated<br>knee<br>extension          | Yes               | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| Standing 3<br>way hip                |                   |                   | Yes                   | Yes               | Yes                   | Yes               |
| Mini wall<br>squats with<br>ball     | Yes               | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| Curl ups<br>with exer-<br>cise ball  | Yes               | Yes               | Yes                   |                   |                       |                   |
| Side walk-<br>ing with<br>resistance |                   |                   | Yes                   | Yes               | Yes                   | Yes               |
| Hamstring<br>stretches               | Yes               | Yes               | D/C to<br>HEP<br>only |                   |                       |                   |
| Balance<br>exercises                 |                   | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| Over<br>ground<br>gait               |                   | Yes               | Yes                   | Yes               | Yes                   | Yes               |
| Obstacle<br>training                 |                   |                   |                       | Yes               | Yes                   | Yes               |
| Stair<br>training                    |                   |                   |                       | Yes               | Yes                   |                   |

Appendix: Interventions provided during each treatment

## Citation

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