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Case Report Open Access

Aquatic and Land-Based Physical Therapy Improves Functional Mobility and Quality of Life in a Patient with Bethlem Myopathy Muscular Dystrophy: A Case Report

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Abstract

We describe a 34-year-old female diagnosed with Bethlem Myopathy Muscular Dystrophy (BMMD). The initial examination revealed decreased muscle strength and range of motion, and impaired gait. The patient was unable to work and lived with her mother who assisted with all activities of daily living. The patient received aquatic and land-based physical therapy interventions 2 times a week for 12 weeks. After 24 sessions, the patient was able to return to work, walk around the park, go shopping and to the movies, participate in the community pool program, and use a fitness center to maintain strength and endurance. Physical therapy was effective in improving safety and independence for functional transfers, mobility, and community participation, which improved patient-reported quality of life. Further research is needed to assess effectiveness of physical therapy interventions on functional mobility, community participation, and quality of life for patients with Bethlem Myopathy Muscular Dystrophy.

Keywords: Limb-girdle muscular dystrophy; Aquatic therapy; Community participation; Interventions; Case-report

Introduction

Muscular dystrophy encompasses a variety of disorders characterized by progressive muscle wasting. Patients with muscular dystrophy are dependent on others for activities of daily living (ADL) and mobility, and report a decreased quality of life (QOL) due to their diagnosis [1]. Two thirds of patients with Bethlem myopathy muscular dystrophy (BMMD), 50 years of age and older, require a wheelchair for community ambulation, and alterations in their home, i.e. elevated toilet seat, grab bars, etc. [2]. At this time, there are no known cures for muscular dystrophy, therefore, current treatments include medical management and physical therapy to address associated impairments and functional limitations.

Limb-girdle muscular dystrophy (LGMD) is comprised of at least 18 different subtypes categorized into Type I, autosomal dominant inheritance, or Type II, autosomal recessive inheritance [3,4]. BMMD, classified as LGMD Type I, has a slow progression and affects the proximal muscles of the body. Impairments caused by BMMD include axial and proximal muscle weakness, shortening of the Achilles tendons, and elbow, spine, and ankle contractures creating difficulty with ambulation and participation in household and community activities [5]. Unfortunately, BMMD is difficult to diagnose due to the onset of symptoms ranging from childhood to adulthood [6], and diagnosis may be difficult due to similarities between BMMD and other neuromuscular disorders.

Physical therapy, early after BMMD diagnosis, is recommended to address associated impairments [6]. Physical therapy is effective to prevent joint contractures, promote walking, and provide patient education regarding mobility aids, splinting, and long-term effects of BMMD [3,4,7]. Resistance training to increase muscle strength in patients with muscular dystrophy is, however, controversial. The underlying mechanism for resistance training is to stress the muscle to cause micro-damage to muscle fibers [8]. This damage stimulates repair and growth adaptation of the muscle to withstand new stresses. In patients with LGMD type II, high and low-intensity resistance exercises demonstrate positive effects of increased muscle strength and endurance, VO_2 max, and work load capacity [9-11]. In patients with fascioscapulohumeral dystrophy, high-intensity cycling activities are effective in increasing muscular strength and endurance without negative effects [9]. However, in patients with Becker muscular dystrophy (BMD) there are conflicting findings with regards to effectiveness of resistance training [9,10]. BMMD impairs the ability of muscle cells to respond normally to external stresses, and therefore, caution is needed when utilizing muscle-strengthening activities as muscle cells may not self-repair and adapt [5].

Aquatic therapy and anti-gravity treadmill training have beneficial effects in the treatment of muscular dystrophy and associated impairments. Aquatic therapy improves balance and coordination, facilitates weight loss and joint protection in obese patients, and improves muscle strength and endurance [12]. The buoyancy of the water decreases the effect of gravity, allowing the patient to perform activities they are unable to perform on land. Patients with Duchenne muscular dystrophy are living longer due to aggressive PT and medical management, and aquatic therapy is increasingly utilized as part of PT management for these patients [13]. In patients with LGMD type II and Becker muscular dystrophy, Berthelsen et al. [14] reported that anti-gravity treadmill training increases 6 minute walk test distance by 8%, improves dynamic postural balance, and decreases steady state HR, without muscle damage. Anti-gravity treadmill training appears to provide a safe and effective intervention to decrease impairments and improve functional mobility in patients with weaknesses cause by muscular dystrophy.

Overall there is little evidence about effectiveness of PT management in BMMD or LGMD Type I diagnosis. The purpose of this case report was to describe the effectiveness of physical therapy on functional mobility, household and community participation, and QOL in a patient with BMMD.

Case report

We describe the case of an obese 34 year-old female with BMMD who sought physical therapy to improve her functional mobility. As a child she exhibited signs of muscular dystrophy and underwent bilateral Achilles tendon releases. She, however, was not diagnosed with BMMD until 18 years of age. Upon initial evaluation, there was a history of falls with significant increase over the past few years. A fall six months prior resulted in a left proximal humerus fracture. Prior to this fall, the patient was living independently in her first floor apartment, working part-time as a teacher, and ambulating without an assistive device. However, subsequent to the fracture, she reported decreased bilateral lower extremity (BLE) strength and endurance, balance, and functional mobility due to six months of inactivity. She now lived with her mother who provided assistance with all ADLs, and was unable to work.

Physical therapy evaluation

Examination of bilateral upper and lower extremities (BUE/LE) revealed intact sensation, diminished reflexes, decreased active and passive ankle and elbow range of motion (ROM), and decreased BUE/LE strength with hip flexors and dorsiflexors being the most impaired (Table 1).

	Left Initial	**Right Initial**
Lower Extremities		
Hip Flexion	0/5	2+/5
Hip Abduction	3+/5	3+/5
Knee Flexion	4-/5	4/5
Knee Extension	3+/5	4/5
Ankle Dorsiflexion	2+/5	2+/5
Upper Extremities		
Shoulder Flexion	3/5	3+/5
Shoulder Abduction	3/5	3/5
Elbow Flexion	4-/5	4-/5
Elbow Extension	3/5	3+/5
Grip Strength	WFL	WFL

Note: Manual Muscle Test 5-point grading scale; 0: no muscle contraction, 2: full AROM through gravity eliminated position, 3: full AROM against gravity, unable to hold against resistance, 4: full AROM against gravity, holds against moderate resistance, 5: full AROM against gravity, holds against maximal resistance. WFL=within functional limits

Table 1: Upper and Lower Extremity Manual Muscle Test Grades.

Examination of gait revealed decreased LLE weight bearing, decreased L hip flexion during swing phase with LLE circumduction, decreased R step length, decreased gait speed, and a wide base of support. The patient also placed her walking stick under her chin during RLE swing phase to assist with LLE weight bearing.

The combination of impairments resulted in a loss of independence in transfers and functional mobility, and increased time needed to perform all activities including: going to the movies, eating dinner with friends, and shopping with her mom. She was also unable to work due to the increased number of falls

Physical therapy plan of care

Patient goals included energy conservation techniques, independence with transfers, and an independent HEP program. Plan of care included aquatic physical therapy 1x/week and land-based physical therapy 1x/week for 8-12 weeks to include gait training, therapeutic activity, therapeutic exercise, neuromuscular re-education and balance, orthotic management and training, and aquatic therapy to achieve patient and physical therapy goals. Due to the patient's obesity and disease process, prognosis to reach patient goals was fair.

Interventions

The initial treatment session included an assessment for an appropriate assistive device for ambulation. Ambulation with a rolling walker was unsafe due to bilateral elbow contractures and poor balance. Using an Arjo sling and walking frame, the patient ambulated 50' in 15 minutes with moderate assistance x2. A goal of ambulation greater than 50 was deemed not appropriate due to decreased balance, disease progression, and energy demands. Mobility goals were modified to include independent use of a power scooter on variety of surfaces for community mobility and return to work. Patient education included disease process of muscular dystrophy, muscle-wasting concepts, possible inability to increase muscle strength, and importance of energy conservation techniques.

Aquatic therapy was initiated during the second week of treatment to increase balance, normalize gait pattern, and improve endurance. During the initial aquatic treatment, the patient exhibited difficulty with foot contact with the bottom of the pool due to increased buoyance in the water, and difficulty pushing a barbell underwater, specifically with her RUE. Leg swings and bicycling in the parallel bars were performed without difficulty. Patient reported fatigue after the first pool session, but increased confidence due to her ability to perform tasks that she was unable to complete on land. In subsequent treatment sessions the patient exhibited improved balance, increased endurance, less fatigue, and was able to exit the pool more quickly. Pool exercises included: (1) step-ups in parallel bars using a 2” step for balance training, (2) progression of UE exercises, and (3) side stepping and forward/backward gait training in chest deep water with focus on normalizing gait pattern in forward direction. Patient also tolerated ten minutes of gait and endurance training using an underwater treadmill at 0.5 mph. The patient was interested in continuing pool exercises as part of a community pool program after discharge. A pool home exercise program was designed for this purpose.

Land-based physical therapy emphasized energy conservation techniques and providing resources to assist with ambulation, ADLs, IADLs, and community related activities. The patient purchased a Pride Go-Go Ultra X scooter (Pride Mobility Products Corp. Exeter, PA, USA) and car lift ramp; patient education included scooter assembly and disassembly, and safe scooter transfers. An orthotist designed a right foot arch support and left ankle foot orthosis (LAFO). The LAFO was fabricated with a hinged ankle clevis joint, 90° down stop, and removable heel build up to provide ankle support, prevent toe drop, decrease plantar flexor contracture, and increase weight bearing on the LLE. The patient was taught to don, doff, and adjust the LAFO, and the importance of using it during ambulation to normalize her gait pattern and increase safety. She wore the LAFO and R foot orthotic during transfer and gait training. Subsequent physical therapy sessions focused on bed mobility and transfer training to increase safety and decrease time required.

Prior to discharge, the patient was instructed in a variety of BUE lightweight, low repetition exercises, and pelvic tilts for core stabilization on a stability ball. She was also instructed in upper body ergometer and stationary recumbent bike cycling for warm up, cool down, and cardiovascular training. Training occurred at a local fitness center by the physical therapist and patient was advised to allow at least a day recovery between exercise sessions. Patient educated to call the doctor or therapist with any problems or questions.

Results

Four weeks after initiating physical therapy, the patient returned to work using a scooter and car lift, and was independent with all scooter transfers. After 12 weeks of physical therapy, the patient was able to walk with a normalized gait pattern using her orthotics and walking stick, and no longer placed the stick under her chin for weight bearing. Using her scooter, she could shop and go to the movies with friends and family members. Lastly, she began to participate in the community pool program, and use the fitness center to maintain strength and endurance. The patient reported less fatigue and more confidence in performing bed mobility, transfers, and ambulation. The patient's progress is illustrated in Table 2.

Task	Initial Evaluation	Discharge
Bed mobility	Min A × 1	Mod I with AD
Transfer Supine-Sit	Min A × 1	Mod I with AD
Transfer Sit-Stand	Min A × 1 (high surface) Unable (low surface)	Mod I with AD (high surface) Min A × 1 (low surface)
Gait Distance/Time	50 feet/8 minutes	100 feet/5 minutes
Gait Assistive Devices	Walking stick used under chin	Walking stick in R hand LAFO, R orthotic
Gait Assistance	Mod A × 2	Mod I with ADs
Work	Unable	Full Time w/scooter
Movies, Shopping, etc.	Unable	Mod I w/scooter

Note: Min A=minimal assistance (≤ 25% assist), Mod A=moderate assistance (>25% to <75% assistance), Mod I=modified independence, AD=assistive device, L=left, AFO=ankle foot orthosis, R=right

Table 2: Outcome Measures.

Discussion

There is little evidence correlating the effects of physical therapy interventions with functional mobility and quality of life in patients with BMMD, or any form of LGMD Type I. This case report illustrates that physical therapy interventions were effective in improving independence in transfers, functional mobility, and community participation that enabled return to work and improved quality of life in a patient with BMMD.

Our patient demonstrated improved balance and endurance after initiating aquatic therapy. These findings concur with Becker's [12] findings that aquatic therapy is beneficial for patients at risk for falling, and increases patient coordination and balance. Given the patient's obesity, the aquatic setting also provided joint protection during exercise. Additionally, the patient reported increased endurance after initiation of resistance training at the local fitness center. These outcomes correlate with the findings of Sveen et al. [10] showing increased muscular endurance following initiation of low-intensity resistance training in patients with BMD and LGMD Type II. Our outcomes suggest that combined aquatic and land-based physical therapy interventions are effective in optimizing functional mobility in patients with BMMD.

Conclusion

Early initiation of physical therapy for patients with LGMD is vital to minimize contractures and promote walking [3,6,7]. Wagner et al. [13] found that in patients with DMD, overall life span improves in patients who participate in aggressive physical therapy and medical management quickly following initial diagnosis. Unfortunately, in this case report, physical therapy was not initiated until sixteen years after initial diagnosis, resulting in multiple impairments causing difficulty with transfers, ambulation, functional mobility, and a poor QOL. Conclusively the management of muscular dystrophies requires a multidisciplinary approach that is initiated as soon as possible following initial diagnosis.

Limitations of this case report include co-morbidities of obesity and amount of time from initial diagnosis to intervention; both are factors that may have impacted response to therapy and outcomes. Additionally combinations of interventions including aquatic and land-based techniques were implemented. Due to this multi-faceted treatment approach, the contribution of a specific intervention to outcomes is unclear. Further research is warranted to assess effectiveness of specific physical therapy interventions on functional mobility, participation, and QOL for patients with BMMD and other forms of LGMD Type I.

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