ORIGINAL ARTICLE

Functional Impairment in Patients with Sporadic Inclusion Body Myositis

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ABSTRACT: ***Introduction:*** We conducted a retrospective chart review of 53 patients diagnosed with sporadic Inclusion Body Myositis (sIBM) who have been followed at the McMaster Neuromuscular Clinic since 1996. ***Objectives:*** We reviewed patient medical histories in order to compare our findings with similar cohorts, and analyzed quantitative strength data to determine functionality in guiding decisions related to gait assistive devices. ***Methods:*** Patient information was acquired through retrospective clinic chart review. ***Results:*** Our study found knee extension strength decreased significantly as patients transitioned to using more supportive gait assistive devices ($P < 0.05$). A decline to below 30 Nm was particularly indicative of the need for a preliminary device (i.e. cane)($P < 0.05$). Falls and fear of falling poses a significant threat to patient physical well-being. The prevalence of dysphagia increased as patients required more supportive gait devices, and finally a significant negative correlation was found between time after onset and creatine kinase (CK) levels ($P < 0.01$). ***Conclusion:*** This study supports that knee extension strength may be a useful tool in advising patients concerning ambulatory assistance. Further investigations concerning gait assistive device use and patient history of falling would be beneficial in preventing future falls and improving long-term patient outcomes.

RÉSUMÉ: Déficits fonctionnels chez les patients présentant une myosite à corps d'inclusion sporadique. ***Contexte :*** Nous avons effectué une revue rétrospective des dossiers de 53 patients chez qui un diagnostic de myosite sporadique à corps d'inclusion (MSCI) avait été posé et qui ont été suivis à la McMaster Neuromuscular Clinic depuis 1996. ***Objectifs :*** nous avons revu l'histoire médicale des patients afin de comparer nos observations à celles faites chez des cohortes similaires et d'analyser les données quantitatives de force musculaire pour déterminer la fonctionnalité afin d'éclairer les décisions prises concernant les appareils d'aide à la locomotion. ***Méthodes :*** Nous avons recueilli l'information au moyen d'une revue rétrospective des dossiers cliniques de ces patients. ***Résultats :*** Nous avons constaté que la force d'extension du genou diminuait significativement à mesure que les patients utilisaient des appareils d'aide à la locomotion offrant plus de support ($p = 0{,}05$). Un déclin à moins de 30 Nm était particulièrement indicatif du besoin d'un premier appareil, soit une canne ($p = 0{,}05$). Les chutes et la crainte de tomber sont une menace au bien-être physique des patients. La prévalence de la dysphagie augmentait à mesure que les patients avaient besoin d'appareils offrant davantage de support à la locomotion et finalement il existait une corrélation négative significative entre le début de la maladie et le taux de créatine kinase (CK) ($p < 0{,}01$). ***Conclusion :*** Ces observations sont en faveur de l'utilisation de la force d'extension au niveau du genou comme outil pour conseiller les patients au sujet des appareils d'aide à la locomotion. Des études plus poussées sur les appareils d'aide à la locomotion et l'histoire de chutes du patient pourraient être utiles dans la prévention des chutes et l'amélioration de l'issue à long terme pour le patient.

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Sporadic Inclusion Body Myositis (sIBM), whose distinctive pathology was first noted by Chou in 1968, has since become known as the most common idiopathic inflammatory myopathy in adults over the age of 50[1,2]. Although the cause of the disease remains unknown, studies support the concurrent existence of autoimmune and degenerative mechanisms[3]. It is a slowly progressive degenerative disorder and can frequently be distinguished from other myopathies by its unique clinical symptoms of muscle weakness particularly affecting the knee extensors, wrist flexors and finger flexors[4]. Dysphagia has also been noted to affect 20% to 80% of sIBM patients[4-9], a broad range that may be partly attributed to the inconsistent questioning of patients, specifically for problems related to swallowing[6]. The diagnosis of sIBM is based on patient's history, electromyography (EMG) findings of a mixed myopathic and neurogenic pattern, elevated creatine kinase (CK) activity, and a muscle biopsy indicating vacuolated muscle fibers, mononuclear T-cells invading non-necrotic fibers, and intra-vacuolar inclusions containing amyloid and other protein deposits[2,3,5].

There is currently no effective treatment for sIBM and management is largely supportive. Clinical trials involving prednisone, intravenous immunoglobin and oral simvastatin have been carried out with little or no positive outcomes[10-13]. The prognosis for sIBM patients is progressive muscle weakness leading to finger weakness, pinch/grip difficulty and knee extensor weakness eventually necessitating gait assistive devices, often stemming from the danger imposed by frequent

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falls that many patients experience in the later years of the disease[14]. Falls are most commonly reported to arise from unprovoked knee-buckling[5]. A recent study found that the time between onset of symptoms and the need for a cane/walking stick was approximately 11 years (y), and for a wheelchair was approximately 16 y[15]. Scales quantifying functional impairment, such as the Inclusion Body Myositis Functional Rating Scale (IBMFRS), have been previously developed in an effort to provide means of tracking the disease's progression[16]. Functional scales have been shown to be sensitive to detecting small changes in function, as well as being efficient and easy to administer[16]. Despite the physically disabling impacts of the disorder, sIBM does not appear to impact a patient's average lifespan[16,17].

The main objective of this retrospective chart study was to determine whether quantitative isometric knee extension strength was associated with a need for gait assistive devices. Secondary objectives were to investigate the relationships between dysphagia and muscle weakness, CK levels and progression of the disease and to explore circumstances surrounding falling in sIBM patients.

Methods

Data was obtained through a retrospective chart review of 53 sIBM patients attending McMaster's Neuromuscular and Neurometabolic Clinic between 1996 and February of 2013. Approval for retrospective chart review was obtained through the Research Ethics Board at the Hamilton Health Sciences Corporation (REB# 13-202-C). The diagnosis was confirmed in all patients by: quadriceps muscle atrophy and weakness, EMG showing a mixed myopathic and neurogenic pattern, and a muscle biopsy showing mononuclear T-cell infiltration of non-necrotic cells plus at least one of rimmed vacuoles (modified Gomori trichrome) and/or intra-muscular 15–18 nm tubulo-filamentous inclusions on electron microscopy (EM).

All patients were diagnosed and evaluated yearly or bi-yearly by one of the authors (MT). At each visit patients were questioned specifically on whether they had difficulty swallowing liquids or solids, or experienced coughing episodes during or after eating. If dysphagia was indicated, patients would undergo a videofluoroscopic swallowing study (VFSS), ranking their dysphagia as mild, moderate or severe. Patients were also questioned concerning their history of falls, the circumstances around the falls, and difficulty with activities of daily living, such as climbing stairs, rising from a chair, getting on and off of the toilet. Finally, the physician questioned patients regarding their use of gait assistive devices including the use of canes, ankle-foot orthoses (AFO), trekking poles, walkers, wheelchairs or scooters, and difficulty with grip strength or fine motor skills using the deep finger flexor, such as manipulation of buttons, belt buckles, etc.

After a detailed history and physical examination all patients underwent quantitative lower isometric knee extension muscle strength testing of the right leg using an isokinetic dynamometer (Biodex System 3, Biodex Medical Systems Inc., Shirley NY), as previously described[18,19]. Hand-grip strength of the right and left hands was completed using a Jamar hand-grip dynamometer (Bollingbrook, New York). Details concerning protocols for both tests are described in Crane, MacNeil, & Tarnopolsky's 2012 article 'Long-term Aerobic Exercise is associated with greater muscle strength throughout the life span'[20]. All strength testing was completed by the same certified kinesiologist.

In order to compare quantitative strength test values to the functional gait abilities of patients, a scale was created to classify patients based on their current methods of gait assistance (Table 1). The gait assistive device scale ranged from 0 to 8 and increased with a patient's growing need for ambulatory assistance. The most widely noted progression in past studies was the transition from non-dependence to reliance on a cane or walking stick, followed by a final reliance on a wheelchair[15,16]. However, such a progression does not encompass ways to differentiate between the levels to which the patient relied on the device in use, and thus the scale was expanded beyond these three transition stages to make such differentiations more clear (Table 1).

Statistical analysis was completed using Statistica and GraphPad Prism Version 6.0 statistical software. Analysis regarding the rate of strength loss over time was completed using

Table 1: Scale classifying patients according to gait assistive device use

Scale	Gait Assistive Device in Use
0	No gait devices are required
1	One gait device (cane, trekking poles, AFO, walker) is suggested by the physician or used occasionally
2	One gait device (cane, trekking poles, AFOs) is used all of the time
3	A combination of cane, trekking poles and/or AFOs is used all of the time
4	A combination of cane, trekking poles and/or AFOs is used all of the time, accompanied by the use of a walker/wheelchair
5	A walker is used full time, no use of a wheelchair
6	A walker is used full time, occasional use of a wheelchair/scooter (e.g. for long distances)
7	A wheelchair/scooter is most prominently used, but gait is still possible with other devices
8	The patient is fully wheelchair bound

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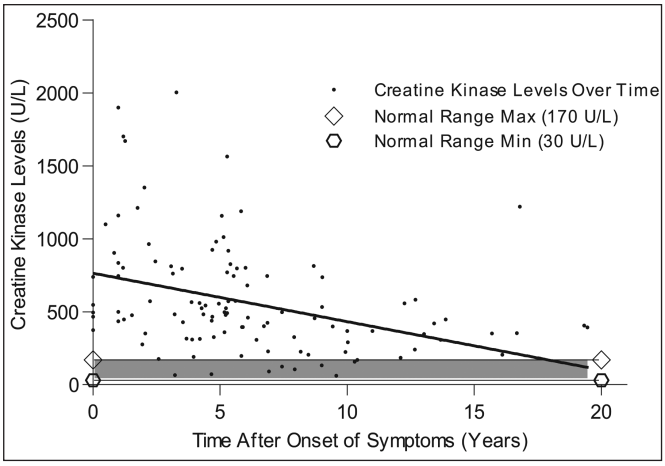

Figure 1: *The negative correlation observed between creatine kinase levels (U/L) in 43 patients and the progression of the disease in years (P < 0.0001). The grey area depicts the normal range of creatine kinase levels (30 to 170 U/L).*

repeated measures analysis of variance (ANOVA). Analysis regarding knee extension strength data of patients in relation to gait device use also utilized repeated measures of AVOVA, as well as unpaired t-tests. A linear regression model was employed to determine the existence of a significant correlation between CK levels and time after onset of symptoms. Correlations between variables were determined using a Pearson correlation co-efficient (r). P-values less than 0.05 using two-tailed testing were deemed significant. All values in graphs and tables are presented as mean +/- SEM.

Results

Patient General Information

Patient demographics and additional information on CK and creatinine levels are summarized in Table 2. Among the 53 patients, 40 were male (75.5%) and 13 were female (24.5%), a distribution (3:1) similar to the commonly reported male-to-female ratios of 2:1 and 3:1[6,11,21,22]. The average age of onset was 60.9 ± 1.34 y, as expected[6]. The average delay between onset of symptoms and diagnosis was 5.5 ± 0.6 y, corresponding to the common five to eight year delay seen in patients with sIBM, often attributed to its slowly progressive nature[13,21]. The average CK levels at diagnosis ranged from normal (55 to 170 U/L for men and 30 to 135 U/L for women[23]) to around 20 times higher than normal. A statistically significant negative correlation between CK levels and time after onset of the disease was found (Figure 1) (r = -0.3832, P < 0.0001). No significant correlations were discovered between creatinine levels and time after onset of symptoms. No unexpected differences were seen between males and females in any of these categories.

Progression of Muscle Strength

Average strength declined in patients over the period of one year and five years after diagnosis (Figure 2). Male and female muscle strength progressions were separated within the figure so as to be comparable to the expected gender-specific strength progressions. No significant differences were seen in strength decline between genders (Figure 2). The average one-year percent muscle decline in patients irrespective of gender was 3.5 (+/- 3.0)%/y for right handgrip in 28 patients and 7.2 (+/-3.7)%/y for left handgrip in 27 patients. Annual percent decline in knee extension for 27 patients was significantly higher than right handgrip at 21.8 (+/- 3.7)%/y (P < 0.01), but not significantly higher when compared to left handgrip (ANOVA). The average five-year percent decline in 18 patients was 27.5 (+/- 4.9)%/5y for right handgrip and 26.6 (+/- 4.2)%/5y for left handgrip. Strength percent decline of knee extension over five years was significantly higher when compared to both handgrip declines at 68.6 (+/- 5.3)%/5y (P < 0.001). Comparisons of right to left handgrip strength percent declines were not significantly different. For gender-based values, refer to Figure 2.

History of Falling

In our group, 37 of 49 (75.5 %) patients reported a history of falls when questioned directly. Patients described knee buckling most frequently as a cause of falling with foot drop/toe stubbing as the second most common cause of falls. The falls were usually triggered by an unexpected event such as a change in surface characteristics (carpet transition, step not anticipated, object on the floor), although some could not describe a particular event

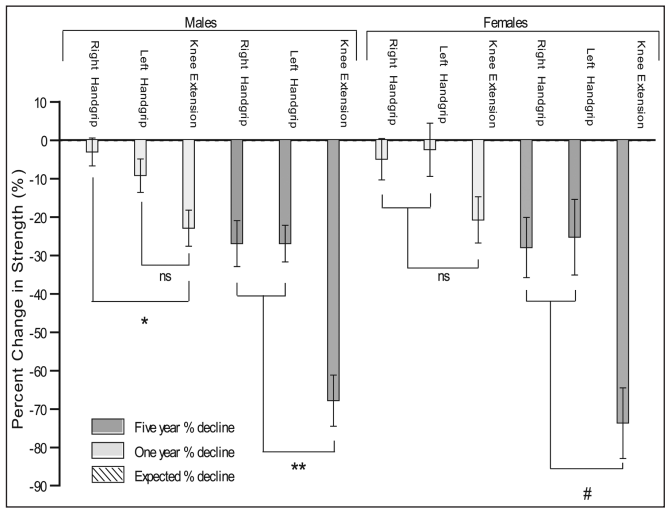

Figure 2: *Average percent change of strength in right handgrip (kg), left handgrip (kg) and knee extension (Nm) as measured over a one-year and a five-year period since diagnosis. Sample size in order from left to right is n = 20, 20, 19, 14, 14, 11, 8, 8, 8, 4, 4, and 3. Error bars represent the SEM. Repeated measures of analysis variance yielded statistically significantly different means (ANOVA) (P < 0.0001). *: p < 0.05, mean strength percent decline in males over one year in knee extension was significantly higher than that of right handgrip. Knee extension strength percent decline over one year was not significantly higher than left handgrip. **: p < 0.001, mean strength percent decline in males over five years was significantly higher than decline of both right and left handgrip. #: p < 0.05 mean strength percent decline in females over five years was significantly higher than decline of handgrip. No significant differences were found when comparing left and right handgrips. No significant differences were found when comparing male and female strength changes in equivalent muscle groups.*

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Table 2: Patient general features

		Number	Mean	Range
Height (cm)	Total	34	176.3 (+/- 1.6)	155 to 193
	Men	27	179.4 (+/- 1.3)	165.1 to 193
	Women	7	164.1 (+/- 2.9)	155 to 177.8
Weight (lbs)	Total	46	181.0 (+/- 5.2)	105 to 247
	Men	35	187.6 (+/- 5.6)	105 to 247
	Women	11	158 (+/- 10.9)	120 to 220.5
Age at Onset of Symptoms (years)	Total	51	60.9 (+/- 1.34)	33 to 76
	Men	39	61.6 (+/- 1.49)	33 to 76
	Women	12	58.8 (+/- 3.02)	40 to 74
Time between Onset of Symptoms and Diagnosis (Years)	Total	50	5.5 (+/- 0.6)	1 to 20
	Men	37	5 (+/- 0.6)	1 to 16
	Women	13	6.7 (+/- 1.6)	1 to 20
Average CK Levels (U/L) at Diagnosis	Total	42	564.1 (+/- 65)	65 to 1900
	Men	31	532.7 (+/- 69)	65 to 1900
	Women	11	652.6 (+/- 156)	132 to 1702
Average Creatinine Levels (umol/L) at Diagnosis	Total	30	68.1 (+/- 3.6)	36 to 111
	Men	24	70.0 (+/- 4.3)	36 to 111
	Women	7	60.1 (+/- 3.8)	41 to 71

Numbers in brackets refer to SEM

other than the "leg just gave out". The mean time between onset of symptoms and the patients' most recent visit was 9.42 ± 0.82 y in those with a positive history of falling episodes, and 3.53 ± 1.06 y in those who denied having ever fallen. Thus, falls seem to become increasingly common as the disease progresses. Among the 37 patients with a history of falls, 11 (29.7%) experienced one or more fractures. The location of fractures varied greatly and included fractures of the wrist, leg, knee, toe, ribs and fingers. Eight patients (20.5%) suffered head injuries, with one resulting in the need for a craniotomy. Although the fear of falling (FOF) was not directly questioned at each visit, four patients specifically expressed anxiety regarding a fear of falling, which has since led them to use wheelchair/scooter full time or to cease certain daily activities.

Gait Assistive Devices and Strength Testing

The retrospective chart review revealed that 48 of the 53 patients' charts held descriptive information concerning their gait assistive device use history. Based on the most recent visits, 16.7% used no gait devices, 36.7% used a cane, trekking pole, or AFO, 28.6% used walkers, and 16.7% were in a wheelchair or scooter. Among the 35 patients using gait assistance, 40% required their first device within five years, 28.6% within five to ten years and 31.4% after ten years of symptoms. Among the eight patients in wheelchairs, one patient required a wheelchair within five years of onset, three patients required a wheelchair within five to ten years, and half required a wheelchair after ten years.

Thorough questioning concerning patients' need for gait assistive devices at clinical visits corresponded to the same dates of quantitative strength testing. Patients' current use of gait assistive devices allowed them to be classified according to the scale in Table 1. Quantitative strength testing, specifically knee extension (Nm), was then compared to each category in the scale. Significant decreases in knee extension were observed as patients moved increasingly through scale categories (Figure 3). Most notably, there was a significant decrease between patients independent of ambulatory devices and those in need of or using a cane/AFO/trekking pole ($P < 0.05$) (ANOVA). Though the test did not yield significance within categories, P-values were lower for patients using primarily walkers and wheelchairs ($P < 0.001$) in comparison to those mainly using canes ($P < 0.01$). Further analysis was conducted through an unpaired t-test, which demonstrated significant differences between patients in need of or using a cane/AFO/trekking pole and those who had transitioned to using walker ($P < 0.05$), and between patients occasionally/constantly using walker and those primarily using a wheelchair ($P < 0.05$) (unpaired t-test).

Swallowing Problems

In our cohort of 53 patients, 31 (58.5%) described having symptoms of swallowing difficulties, whereas 22 (41.5%) had not reported any swallowing issues at the time of the last clinic visit. Amid those affected, 86.7% developed swallowing issues after the onset of other symptoms, such as muscle weakness of the quadriceps or finger flexors, the average time from onset to swallowing issues being 8.12 +/- 1.16 y. Four patients (13.3%) described swallowing issues as among the first symptoms experienced.

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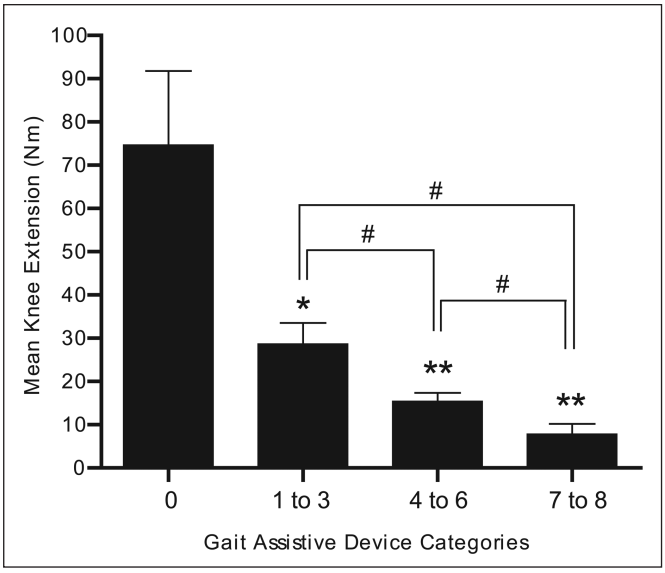

Figure 3: *Mean knee extension strengths (Nm) (mean +/- SEM) of 48 patients categorized by the scale in Table 1. Sample size in order from left to right is n = 9, 21, 19, and 7. Repeated measures of analysis variance (ANOVA) demonstrated statistically significant means (P < 0.0001). *: p < 0.01, means of patients in need of or using cane/AFO/trekking pole were significantly lower than those independent of devices. **: p < 0.001, means of patients using a walker for some/all distances and of patients predominantly using a wheelchair were significantly lower than those independent of devices. No other significant differences were found between categories according to the ANOVA test. Additionally significant differences between categories were found, however, when an unpaired t-test was conducted (#: p < 0.05).*

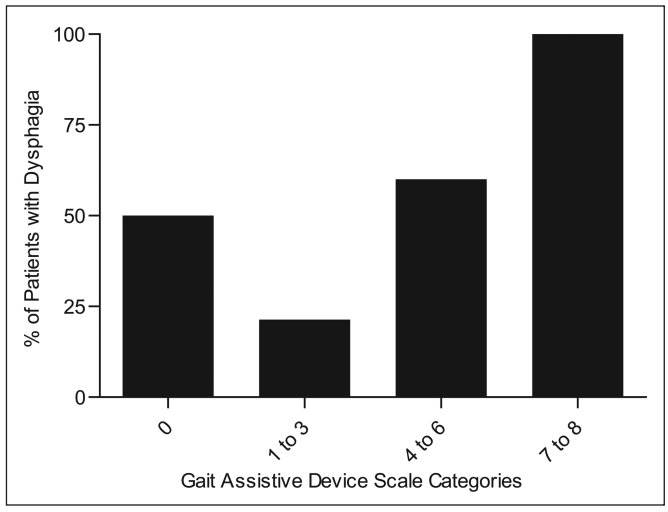

Figure 4: *Percent of patients experiencing dysphagia categorized by the scale in Table 1, specifically depending on if they were independent of all types of gait devices (0), in need of or using a cane/AFO/trekking (1 to 3), using a walker occasionally or full time (4 to 6) or using primarily a wheelchair (7 to 8). Actual numbers of patients suffering from dysphagia in each category were 3 of 6 (0), 3 of 14 (1 to 3), 12 of 20 (4 to 6) and 8 of 8 (7 to 8).*

Swallowing issues were additionally investigated in relation to ambulatory assistance. Patients were divided into four categories depending on their gait assistive device use, and the percentage of patients suffering from dysphagia in each group was calculated. The proportion of patients with swallowing issues appeared to increase in incidence as patients became more reliant on gait devices. Specifically, a ~ 35% increase was seen when comparing patients using primarily a cane to those mainly using a walker, as well as between those using a walker to those in a wheelchair (Figure 4). There was, however, a relatively high percent (50%) of patients independent of ambulatory assistance that suffered from dysphagia (Figure 4).

Eighteen patients (58.1%) with dysphagia underwent a VFSS. Among the four patients in wheelchairs who underwent the study, three were classified as having moderate to moderately severe dysphagia, while the fourth had mild dysphagia. Among patients in walkers, five were classified as having moderate to moderately severe dysphagia, while three had mild dysphagia. Among those using a cane, an AFO or trekking poles, two patients were ranked as having mild dysphagia, whereas one had no visible signs of dysphagia, despite experiencing symptoms. Finally, of the three patients independent of ambulatory assistance receiving the study, one had severe dysphagia, one had moderate dysphagia and one had mild dysphagia.

Discussion

One of the most notable findings of the current study was the significant correlation between lower isometric knee extension strength and increased reliance on ambulatory assistive devices. A study conducted in 2012[21], as well as the IBMFRS outlined in 2008[16], demonstrate that knee extensor strength and functional scale ratings can be potentially predictive of functional outcome; however, the current data is the first to investigate knee extensor strength specifically in relation to gait assistive devices. In general, our data suggest that once knee extension strength declines below ~ 30 Nm it is likely the patient will be requiring or using a cane or trekking pole. A further decline to less than 20 Nm is indicative of a greater degree of support (walker), and knee extension strength approaching 5 Nm indicates wheelchair or scooter dependence. The relationship between an objective/quantitative strength measurement and a significant functional outcome is important for prognostication, patient counseling, as an outcome for clinical trials, and potentially for insurance coverage. Our data are likely to be transferable to the broader IBM population given that the gender ratio, average age of onset, and time between onset of symptoms and diagnosis were similar in our cohort of patients to those reported previously[2,6,13,22].

The average percent decline in muscle strength is another significant tool in predicting and preparing patients for possible future outcomes. The handgrip strength decline that we observed reflected similar values to past report declines of 5-10%/y declines[15]. The average decline in knee extension strength was significantly greater than right and left handgrip strength decline over a five-year period, supporting the repeatedly recognized fact that quadriceps are particularly affected in IBM patients[24,25]. Handgrip and knee extension strength also declined at a higher

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rate than normative and observed rates of controls in previous studies[26-28]. Men and women experienced very similar percent decline in all strength categories, suggesting that sex does not affect strength decline. These findings offer a numerical value to this rapid decline, and may be useful in future clinical trials. Furthermore, the test/re-test coefficient of variation for measuring peak Jamar handgrip strength with our device is < 2%[29], and peak isometric knee extension torque has been found to have a variation of < 5%[30], supporting their reliability for reproducibly capturing small changes in muscle strength. It is important to note that strength test values varied greatly between patients; consequently, it is imperative that future studies use within-subject designs with either a cross-over or a personal natural history run-in period for each participant.

Analyzing the circumstances surrounding falls in sIBM patients is necessary to predict occurrence and identify methods of preventing them. We show that falls are quite harmful as they caused approximately one third of our patients to become seriously injured with either a fracture or head injury. Injuries were not particularly localized to one area of the body. Since falls were often elicited by transitioning to uneven surfaces or steps, and since falling becomes more common with increased muscle weakness, patients should be advised early on to consider stair lifts or modifying their homes to reduce the hazardous surfaces or subtle steps that may result in tripping. A more quantitative approach to examining the frequency and effects of falls in sIBM patients would be beneficial, as this study was limited to a retrospective review of observations recorded qualitatively. It is possible that numbers of falls can be over or under represented in such circumstances.

Another negative consequence of falling is the fear it instills in patients. Fear of falling in the aging population has been described as a factor contributing to a decrease in the quality of life[31]. Although only four patients self reported anxiety regarding FOF, it is likely that we under-estimated the prevalence of this and the negative effect it has on quality of life. Often the transition to more significant gait assistance was triggered by a major fall with or without a fracture. Further research into this subject is needed to determine the prevalence of FOF in sIBM patients and potential ways of diminishing its negative impact on patients' quality of life.

The percentage of sIBM patients reporting dysphagia differs greatly within literature (20-80%); however, recent studies as well as our own indicate reasoning for this variation. It has been reported that dysphagia is indicative of a worsening status[8], and that it is positively correlated with the time following the onset of quadriceps weakness[32]. We have similar findings in that the prevalence of dysphagia was much higher as reliance on more supportive devices increased (Figure 4). Our VFSS findings also indicate a worsening severity in patient's dysphagia as the disease progresses, as the majority of patients in a walker or wheelchair, 75% and 62.5% respectively, were classified as having moderate to moderately severe dysphagia, whereas all patients using canes/AFOs/trekking poles were classified as having mild to no visible dysphagia. However, it is also important to note that dysphagia can be an early and initial symptom, as four independently ambulatory patients (13% of those with swallowing problems) presented with swallowing issues.

Collectively, our data and that previously reported[8,32] supports that dysphagia increases in incidence as muscle weakness increases, which suggests that the wide range of previously reported dysphagia was very likely reliant on the stage of muscle weakness of the patient cohort that is also related to disease duration. Our data suggest that careful questioning for symptoms of dysphagia should be done at annual follow-ups, including questioning the spouse or partner about post-swallowing coughing. Given the universal finding of dysphagia in non-ambulatory patients (8/8), we would suggest that patients with knee extension strength values of < 20 Nm (when they are at high risk of needing a scooter/wheelchair) be sent for quantitative swallowing assessments even if they do not have subjective symptoms.

Finally, numerous past studies have noted abnormal serum CK levels in sIBM patients; however, few have investigated how levels change over time as a function of strength loss, and presumably muscle loss. Surprisingly, Badrising *et al* 2005[32] investigated the relationships between CK levels and age, age of onset, and symptom duration, but found no correlation. In our cohort, we found significantly lower serum CK as the patients' disease progressed ($P < 0.0001$). This is logical, since CK production should decrease as muscle mass decreases. Thus, the patients' recorded CK levels may depend upon the number of years they have been living with the disease. This trend may also partly account for the wide range of average CK levels that have been reported in past studies.

Proper considerations should be made prior to recommending ambulatory assistive devices based solely on the above data, however, as certain limitations should be taken into account. The scale employed in this study is currently not validated, but was developed when a suitable, already existing one could not be found. Scales, such as the modified Gait Efficiency Scale (mGES)[33], were considered, however they were not conducive to the retrospective nature of the study, or to the observed sIBM trend of progression, and would not allow for the appropriate, more specified classifications to be made. Additionally, the scale could not account for all mild differences in dependencies on gait devices, for instance differentiating between patients using a cane twice a week in comparison to those using a cane five days a week. A prospective study is necessary to validate the findings prior to accepting the data as definitive. Finally, we acknowledge that dynamometry techniques are not routinely used in clinical settings, and neither should strength measurements be used as the sole factor in making decisions related to gait assistive device use.

Conclusion

This retrospective chart review resulted in a few notable findings. First, knee extension strength may be clinically useful in advising patients concerning their use of gait assistive devices to prevent falling episodes. Knee extension strength below 30 Nm for a patient independent of gait assistive devices may be reason to advise strongly in favour of a preliminary device, such as a cane or trekking pole. Falling and FOF were shown to have a negative impact on patients' health and further research regarding these topics is required to aid patients in maintaining the highest possible quality of life. Moreover, we postulate that the likelihood a patient will develop dysphagia is not a static

percentage of 20-80%, but is a continuous variable that increases as the patient's muscle weakness/disease duration progresses. Hence, patients should be consistently questioned regarding their swallowing capabilities and educated on the possibility that dysphagia becomes more common as the diseases progresses. Finally, we found a negative correlation between CK levels and time after onset of symptoms, offering an explanation for the varying mean CK levels reported in past studies.

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