Gait patterns of patients with inclusion body myositis

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ABSTRACT

Inclusion body myositis (IBM) is a progressive, inflammatory muscle disease that is known to cause quadriceps weakness and knee buckling during gait. This is the first known report of gait characteristics in patients with IBM. Nine subjects with IBM and quadriceps weakness underwent gait analysis and quantitative strength testing. A wide range of strength and gait abilities were present in the subject group. Subjects with stronger knee extensors exhibited nearly normal sagittal knee kinematics and kinetics. As quadriceps strength decreased, kinematic and kinetic patterns were increasingly abnormal. Exceptions to this pattern could be explained by examining strength at adjacent joints. Gait analysis and strength testing is a helpful tool in evaluating the functional status of this population and aiding in determination of the needs for interventions such as assistive devices.

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1. Introduction

Inclusion body myositis (IBM) is a progressive, inflammatory muscle disease with no known causes and no proven treatment. It affects 5–10 people per million and mostly males [1,2]. IBM is the most common acquired inflammatory myopathy in persons older than 50 years [3–5]. Because of the slowly progressive nature, diagnosis typically does not take place until 6 years after the onset of symptoms [3,6]. Also, IBM is often misdiagnosed as other types of myopathy because of an inconclusive muscle biopsy or pattern of weakness. For this reason, IBM is thought to be under diagnosed [1,7]. Once diagnosed, treatment is usually of the supportive nature rather than curative because IBM does not respond to typical immunosuppressive treatments [3,8,9]. Current treatments aim to slow the rate of progression of the disease rather than cure it [5,10]. True treatment efficacy is difficult to gauge, however, because of the relatively slow progression, rate of progression differing between patients, and the fact that older patients tend to decline more rapidly, although the reason for this is not entirely clear [10,11].

Patients with IBM have a distinctive pattern of weakness. Although IBM can affect both the proximal and distal muscles of the extremities, the disease almost universally involves the wrist and forearm finger flexors, quadriceps, ankle dorsiflexors, and swallowing muscles. Ventral muscle groups are more affected than dorsal and girdle muscles, which can help preserve postural stability and locomotion [12]. Most patients with IBM present with an initial complaint of weakness, predominantly in the proximal lower limbs.

Strength usually decreases 5–10% per year, although rates of progression can vary [3,10,12]. Yet most patients remain ambulatory for many years after diagnosis. However, because the quadriceps are among the most affected muscles with IBM, patients often complain of frequent falls or buckling of the knees. Up to 75% of IBM patients will use an assistive device such as a cane as the disease progresses, with some moving to a wheelchair mainly because of a fear of falling [12]. Three quarters of patients with IBM report frequent falls, and frequency declines for a time after initial diagnosis before increasing again as the disease progresses [12]. Although first described in the late 1960s, IBM remains poorly understood despite increasing interest over the last two decades. Several studies from large neuromuscular centers have been published showing that IBM is not as rare as previously thought, comprising up to 28% of all inflammatory myopathies [1,5,6,13–15]. More recently published reports on IBM concentrate on disease epidemiology or drug trials. No comprehensive description of gait of patients with IBM exists. Therefore, the purpose of this study was to report on gait characteristics of patients with IBM. We hypothesized that as quadriceps strength decreased, the knee moment would progress from an internal extension moment of normal magnitude to a flexion moment, and that gait parameters at adjacent joints would be affected as well. We also expected a statistical correlation between strength and gait parameters.

2. Methods

2.1. Participants

Nine subjects with IBM were enrolled after a screening interview with the principal investigator (THO) (7 males, 2
females, average age 61 ± 9, average BMI 27.2 ± 4.0). Time since onset of symptoms was 6 ± 4 years. All participants signed a written consent form approved by the local Institutional Review Board. All subjects reported quadriceps weakness at enrollment, and four of the nine subjects reported using a cane at times. All subjects were independent in activities of daily living, but had experienced falling or knee buckling in the past.

2.2. Motion analysis testing

Reflective markers were placed on each subject as described by Kadaba et al. [16]. Markers were placed bilaterally on the acromion processes, anterior superior iliac spines (ASIS), lateral femoral condyles, lateral malleoli, the spaces between the first and second metatarsal heads, heels, on 5 cm wands placed at mid-thigh and mid-shank, and on the sacrum. The markers placed at bony prominences were used for establishing local anatomic coordinate systems for the pelvis, thigh, shank, and foot. One trial corresponding to the standing position (static data) were recorded in order to calculate the location of the joint centers. The subjects were asked to walk on a level surface while marker trajectory data was collected at 60 Hz using a 10 camera Real Time Motion Analysis system (EvaRT 5.04, Motion Analysis Corporation, Santa Rosa, CA). Ground reaction force data was sampled simultaneously from four force plates (2 AMTI model BP400600, Advanced Mechanical Technology, Inc., Watertown, MA and 2 Kistler model 9281B, Kistler Instrument Corp., Amherst, NY). All subjects walked in order to calculate the location of the joint centers. The subjects were asked to walk on a level surface while marker trajectory data was collected at 60 Hz using a 10 camera Real Time Motion Analysis system (EvaRT 5.04, Motion Analysis Corporation, Santa Rosa, CA). Ground reaction force data was sampled simultaneously from four force plates (2 AMTI model BP400600, Advanced Mechanical Technology, Inc., Watertown, MA and 2 Kistler model 9281B, Kistler Instrument Corp., Amherst, NY). All subjects walked

2.3. Instrumented strength testing

Instrumented strength testing was performed on a strain-gauge tensiometer muscle testing system following a validated standardized testing method, derived from the Tufts Quantitative Neuromuscular Exam (TQNE) [Avery Medical, LLC, Gainesville, GA] [17]. All strength testing was done by a licensed physical therapist trained in the use of the testing equipment. Isometric strength of the bilateral hip flexors, hip extensors, hip abductors, knee flexors, knee extensors, ankle plantarflexors, and ankle dorsiflexors was measured with the subjects in standardized positions (Table 1). Two trials were performed for each muscle group, with the maximum value recorded. The data was expressed as joint moments by multiplying the measured force by the distance between the joint of interest and position of the strain-gauge testing strap and normalizing by subject height and weight. Subjects were ranked by normalized knee extensor (quadriceps) strength on their weaker side as determined by knee extensor strength.

2.4. Statistical methods

Correlations between gait parameters and strength data were determined using Pearson correlation coefficients. Statistical significance was set at p = 0.05.

3. Results

A range in gait patterns was evident within this group when examining the gait data from the weaker limb (Fig. 1), especially knee kinematics and kinetics and ankle kinematics. Some of the subjects had nearly normal sagittal knee motion, complete with a loading response in early stance and neutral positioning in late stance. However, most subjects maintained an abnormal hyper-extended knee position throughout stance. All participants had a normal peak knee flexion during swing, with a few having excessive peak knee flexion. A similar trend was seen in the sagittal knee moment. About one-third of the subjects had a normal internal extension moment peak in early stance, while the remainder of the participants showed an internal flexion moment throughout stance. All participants had a decreased peak knee moment compared to normal. Ankle dorsiflexion also varied. About one-third of the subjects had a nearly normal dorsiflexion curve, with the remainder having increased plantarflexion in early stance and a lack of dorsiflexion in swing.

Strength values for the weaker limb for all muscles tested also showed a wide range (Table 2), which had a direct effect on gait. All subjects were weak when compared to age and gender matched normals in the literature [18], with the weaker limb knee extensor strength less than 40% of expected values for all subjects. The subject with the strongest quadriceps, Subject #3, had nearly normal kinematic and kinetic patterns (Fig. 2). As expected, kinematic and kinetic abnormalities increased with decreasing strength, particularly in knee position during stance, sagittal knee moment, and ankle dorsiflexion (Fig. 2). In general, with decreasing strength, knee position went from a normal, slightly flexed position during loading response to a progressively more hyperextended position throughout stance. Similarly, the sagittal knee moment showed a normal pattern but slightly decreased magnitude with the stronger subjects, becoming more flat, and finally an internal flexion moment as opposed to the normal internal extension moment. This indicates a change in patient approach to stability during stance, with the stronger subjects relying on their available

<table>
<thead>
<tr>
<th>Muscle group</th>
<th>Patient position</th>
<th>Limb position</th>
<th>Measurement position</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip extension</td>
<td>Supine with trunk supported by wedge (at 20° of hip flexion)</td>
<td>Hip at 20°, knee in full extension</td>
<td>Distal thigh</td>
</tr>
<tr>
<td>Hip flexion</td>
<td>Supine with trunk supported by wedge (at 20° of hip flexion)</td>
<td>(opposite hip and knee at 90°)</td>
<td>Distal thigh</td>
</tr>
<tr>
<td>Hip abduction</td>
<td>Supine</td>
<td>Hip at 20°, knee in full extension (opposite hip and knee at 90°)</td>
<td>Distal thigh</td>
</tr>
<tr>
<td>Knee extension</td>
<td>Sitting upright; rolled towel under distal thigh</td>
<td>Hip and knee extended (opposite hip and knee at 90°)</td>
<td>Proximal to lateral malleolus</td>
</tr>
<tr>
<td>Knee flexion</td>
<td>Sitting upright; rolled towel under distal thigh</td>
<td>Hip and knee 90°</td>
<td>Proximal to lateral malleolus</td>
</tr>
<tr>
<td>Ankle plantarflexion</td>
<td>Supine</td>
<td>Hip and knee extended</td>
<td>Metatarsal heads</td>
</tr>
<tr>
<td>Ankle dorsiflexion</td>
<td>Supine</td>
<td>Hip and knee extended</td>
<td>Metatarsal heads</td>
</tr>
</tbody>
</table>

muscle strength and the weaker subjects taking advantage of mechanical stability via body positioning. Ankle dorsiflexion also went from nearly normal to an increasing amount of plantarflexion in stance and decreasing dorsiflexion in swing. When looking at the strength data, the subject with the 7th best knee extensor strength, Subject #6, seems to be a cutoff for where this change takes place (Fig. 2, panes d–f), especially at the knee. At this strength level, the knee moment curve does not display a normal internal knee extension moment, and the knee position during stance also changes from the slight flexion during loading response to the inherently stable hyperextension. 

The gait data from this group does have exceptions to the general pattern, however. One of the subjects with the best strength, Subject #5, was unexpectedly hyperextended at the knee

Table 2
Isometric strength for the side with weaker knee extensors.

<table>
<thead>
<tr>
<th>Subject</th>
<th>Hip flexion</th>
<th>Hip extension</th>
<th>Hip abduction</th>
<th>Knee flexion</th>
<th>Knee extension</th>
<th>Ankle plantarflexion</th>
<th>Ankle dorsiflexion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4.9621</td>
<td>3.2752</td>
<td>7.3562</td>
<td>1.7347</td>
<td>0.3021</td>
<td>Not Tested</td>
<td>0.3354</td>
</tr>
<tr>
<td>2</td>
<td>3.9598</td>
<td>5.0992</td>
<td>6.6743</td>
<td>3.6277</td>
<td>1.6714</td>
<td>2.7519</td>
<td>0.1733</td>
</tr>
<tr>
<td>3</td>
<td>8.9635</td>
<td>13.1306</td>
<td>9.5663</td>
<td>3.5585</td>
<td>4.8525</td>
<td>2.8583</td>
<td>1.9605</td>
</tr>
<tr>
<td>4</td>
<td>7.3839</td>
<td>8.6709</td>
<td>6.6402</td>
<td>2.9927</td>
<td>2.3634</td>
<td>2.3526</td>
<td>0.3229</td>
</tr>
<tr>
<td>5</td>
<td>5.5405</td>
<td>7.7657</td>
<td>8.0592</td>
<td>4.6860</td>
<td>4.1296</td>
<td>7.4000</td>
<td>1.3304</td>
</tr>
<tr>
<td>6</td>
<td>4.9962</td>
<td>12.2539</td>
<td>6.5635</td>
<td>3.5222</td>
<td>1.3352</td>
<td>7.4000</td>
<td>1.3304</td>
</tr>
<tr>
<td>8</td>
<td>8.7926</td>
<td>8.9105</td>
<td>10.3009</td>
<td>2.7512</td>
<td>2.8815</td>
<td>4.6992</td>
<td>2.0687</td>
</tr>
<tr>
<td>9</td>
<td>5.6170</td>
<td>6.0656</td>
<td>6.1072</td>
<td>2.1524</td>
<td>3.0291</td>
<td>3.9823</td>
<td>1.7972</td>
</tr>
</tbody>
</table>

Values shown are unitless and expressed as % body weight * height [torque (N m) normalized to subject height (m) and weight (N)].

Fig. 1. Knee sagittal kinematics (a) and kinetics (b) and ankle kinematics (c) for patients with IBM. The solid black lines represent individual subjects. The grey band represents normal ± 1 SD.

Fig. 2. Knee sagittal kinematics (a) and kinetics (b) and ankle kinematics (c) for the strongest subject, Subject #3. All closely follow the normal band. Knee sagittal kinematics (d) and kinetics (e) and ankle kinematics (f) for the subject with the 7th best knee extensor strength, Subject #6. Knee sagittal kinematics (g) and kinetics (h) and ankle kinematics (i) for the subject with the weakest knee extensors, Subject #1.

Ankle kinematics are less abnormal than in other weak subjects. An excessive plantarflexion in stance. Knee sagittal kinematics (d) and kinetics (e) and ankle kinematics (f) for Subject #7. Even though one of the weakest subjects in knee extensor strength, the moment curve is less abnormal and knee position during stance is near neutral as opposed to the more hyperextended position seen in other weaker subjects. Ankle kinematics are less abnormal than in other weak subjects.

There were notable exceptions to the general pattern of kinematics and kinetics, however. A subject with less knee extensor weakness showed unexpectedly abnormal knee kinematics and kinetics with increased weakness at the ankle. Conversely, a subject with more quadriceps weakness showed surprisingly normal gait parameters because of less weakness at the ankle.

Many investigators have suggested that knee extensor strength and knee gait mechanics cannot be taken in isolation, since causes of or compensations for knee abnormalities can occur at neighboring joints. Over 30 years ago, Sutherland demonstrated how simulated ankle weakness affected knee position [19]. In her textbook on human movement, Perry discussed at length the possible links between abnormal knee kinematics and ankle strength [20]. More recently, modeling by Goldberg showed that ankle plantarflexors can provide compensation for weakness in many lower extremity muscle groups [21], and induced accelerations work by Siegel added that the exact compensatory strategy for knee weakness varies with adjacent muscle weakness [22]. The kinematic patterns and correlations between strength and gait parameters at adjacent joints found in this study support these observations. Knee extensor weakness in patients with IBM has a general pattern, but if a subject has relatively good ankle or hip strength, as is often the case with this unique disease, patients can still walk well.

This study has several limitations. IBM is a relatively rare disease, and due to geographic limitations on recruitment, only nine subjects were studied. Because the exact progression timeline of IBM is not fully known, and because of the delay often seen in diagnosing this disease, we cannot be sure where in the disease process these participants were. Therefore this may not have been an accurate depiction of all stages of the disease, especially those in the early stages. Regardless, a strength of this study is that the participants had a varied set of functional abilities and strengths. The gait patterns within this small group are applicable to the IBM population as a whole. Further, the gait patterns observed in this disease provide valuable insight into the gait of patients with other disorders where knee extensor weakness is a chief complaint.

4. Discussion

This is the first known report describing gait patterns of persons with IBM. Previous work has shown that IBM has a unique pattern of weakness and progression. The most common complaint is knee buckling and quadriceps weakness. The patient group in this study demonstrates that although all patients with IBM may be classified as weak, there are a wide range of functional abilities. Specifically, there was a range of knee kinematics and kinetics. While the subjects with stronger quadriceps had more normal knee kinematics and kinetics, those with the weaker knee extensors tended to walk with a more hyperextended knee during stance and an internal knee flexion moment, indicating the force vector remained in front of the knee joint during all of stance in order to achieve mechanical stability.

There were noted exceptions to the general pattern of kinematics and kinetics, however. A subject with less knee extensor weakness showed unexpectedly abnormal knee kinematics and kinetics with increased weakness at the ankle. Conversely, a subject with more quadriceps weakness showed surprisingly normal gait parameters because of less weakness at the ankle.

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Conflict of interest statement

The authors have no conflict of interest regarding any of the material in the manuscript and meet the criteria for authorship as defined in your submission guidelines.

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