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Compensatory mechanisms during walking in response to muscle weakness in spinal muscular atrophy, type III

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Abstract

Our knowledge on altered neurological control of walking due to weakness of various muscle groups of the lower extremities is limited. The aim of this study was to assess kinematic, kinetic and electromyographic (EMG) walking patterns in a functionally homogeneous group of seven subjects with spinal muscular atrophy, type III (SMA group) and compare them with normal data obtained from nine healthy subjects (CONTROL group) in order to identify characteristic compensatory changes. Muscle strength at the ankle and knee joints was assessed using isokinetic dynamometry to determine variability in muscle strength: this was found to be similar in the two groups. Kinematic, kinetic and EMG patterns were assessed during walking in the SMA and CONTROL groups. The results showed changes in the activity of ankle plantarflexors and associated control of the center of pressure during loading response and midstance, which facilitated minimization of the external flexion moment acting on the knee and hip in the SMA group. Additionally, we identified distinct and consistent changes in the control of hip rotators that act to rapidly extend the hip early in stance phase and in the control of contralateral hip abductors that act delay weight shift onto the leg entering the stance phase. From these results we can conclude that the most important muscle groups compensating for reduced strength in knee and hip muscles are the ankle plantarflexors, hip rotators and hip abductors. This finding would have direct application in rehabilitation treatment programs.

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1. Introduction

Neuromuscular disorders include heterogeneous groups of diseases of the motor unit, with muscular weakness as a common and usually predominant clinical sign. Muscle weakness is the major limiting factor that profoundly influences walking ability in patients with neuromuscular disorders [1,2]. However, clinical data on gait in patients with specific neuromuscular disorders are scarce. Clinical observation of walking in people with neuromuscular disorders suggests that various adaptations may occur [3,4]. In terms of spatio-temporal gait characteristics, coping

responses include decrease of walking speed, step length and swing time in order to reduce the mechanical output requirements of weak muscles. Various kinematic and kinetic adaptations, including forward pelvic tilt with lumbar lordosis, reduced loading response, rapid hip extension following initial contact, equinus foot position and lateral trunk motion were observed. The seminal work of Sutherland et al. [5,6] who investigated the walking of a group of patients with Duchenne muscular dystrophy (DMD) in various phases of disease progression, laid the foundations to our current understanding of the pathomechanics of gait in DMD.

Different neuromuscular disorders exist in which muscle groups may be affected in a rather similar pattern. Spinal muscular atrophy (SMA), for example, is characterized by

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symmetrical muscle weakness affecting proximal muscles mostly, as is DMD [7]. Based on this similarity, a hypothesis that similar compensatory changes in gait could be expected in both pathological conditions is plausible. Recently, Armand et al. [8] investigated kinematic and kinetic gait patterns in DMD and SMA type II by means of instrumented gait analysis, including two patients per group. Their results indicated that distinctive differences in the pathomechanics of walking in the two groups existed which would be in accordance with more distinct distribution of muscle weakness in SMA [9]. However, due to the small number of subjects and low walking speed, their results were inconclusive.

The objective of this study was to examine kinematic, kinetic and dynamic EMG patterns of a larger group of patients with spinal muscular atrophy type III. These patients can usually walk independently. In the early stages of the disease their walking speed is comparable to normal. Our aim was twofold: first, to determine characteristic compensatory mechanisms by comparing them with a group of neurologically intact individuals; second, to qualitatively compare group averaged kinematic and kinetic patterns of the SMA group with a group of DMD patients assessed by Sutherland at a comparable functional stage [5,6]. The aim was to investigate the hypothesis that similar compensatory strategies are used in both pathological conditions.

2. Methods

2.1. Subjects

The experimental group consisted of seven adult subjects (age 39.7 ± 11.04 years, height 169 ± 10 cm and body mass 68 ± 17 kg), four males and three females who were diagnosed with SMA, type III. They were selected from the group of all adult ambulatory subjects (nine subjects) diagnosed with SMA type III who reside in Slovenia. The remaining two subjects were excluded from the study because of an evidently early or progressed phase of the disease as judged by observational clinical gait analysis (evidently too fast or too slow walking) and manual muscle testing of the lower limb muscle strength. In addition, nine healthy individuals with no history of musculoskeletal or neurological disease (age 33.11 ± 2.66 years, height 178 ± 12 cm and body mass 76 ± 14 kg), six males and three females, were included as a control group. The study was approved by the appropriate ethics committee and the subjects provided informed consent.

2.2. Experimental conditions

Muscle strength of the ankle and knee muscles was assessed by means of isokinetic dynamometry in both concentric and eccentric modes of muscle contraction. The assessment was performed while the subjects were in sitting position with first the ankle dorsiflexion/plantarflexion axis (range of motion $15\text{--}25^\circ$) and later knee flexion/extension axis (range of motion $90\text{--}0^\circ$) aligned with BIODEX 3 (Biodex Medical Systems, New York, USA) dynamometer motor axis. The velocity of movement was limited to $60^\circ/\text{s}$ for concentric and eccentric muscle contraction. Each movement was repeated

four times. Hip and trunk muscle strength were not tested due to the inability of patients with SMA to perform the required isokinetic strength assessment.

Subjects were asked to walk across a 10 m-gait laboratory walkway at their preferred speed. A VICON motion capture and analysis system (VICON 370, Oxford Metrics Ltd., Oxford, UK) was used to capture motion of the lower limbs, pelvis and trunk. Reflective markers were attached to the subjects' skin over designated landmarks according to the specifications provided by the manufacturer of the system (Vicon Clinical Manager). Motion data were sampled at 50 Hz. Two AMTI force plates (AMTI OR-6-5-1000, Advanced Mechanical Technology Inc., Watertown, MA) that were positioned in the center of the walkway were used for recording ground reaction forces. Force data were sampled at 1000 Hz. Joint angles, moments and powers were calculated using Vicon Clinical Manager. Electromyographic (EMG) activity of selected muscles (tibialis anterior, soleus, gastrocnemius, rectus femoris, semitendinosus, gluteus medius, rectus abdominis and erector spinae) was assessed by repositionable surface electrodes (3MTM Red Dot™) and amplified by MyoSystem 2000 (Noraxon Inc., USA) at sampling frequency 1000 Hz. EMG data were filtered using a 4th-order low pass Butterworth filter with cut-off frequency of 7 Hz. At least three steps of each leg were captured for analysis.

2.3. Data analysis

Maximal values of isokinetic joint torques were extracted. For each subject the averaged values from four trials were calculated for each mode of muscle contraction and used in subsequent averaging across each group.

Gait velocity, stride length and cadence data were extracted and tested in an independent *t*-test for differences between the two groups. Ankle (dorsiflexion/plantarflexion), knee (flexion/extension) and hip (flexion/extension, adduction/abduction and internal/external rotation) angles, joint moments and powers were calculated. Joint moments and powers were normalized for body mass and reported in Nm/kg and W/kg, respectively. For each subject the averaged values from three trials for each leg were calculated and used in subsequent averaging and statistical analysis of the data for the whole group and for each group separately. Gait cycle terminology as introduced by Perry [10] was adopted to define instants of characteristic peak values of kinematic and kinetic trajectories in the gait cycle.

Similarly, EMG data were averaged in each subject and used in subsequent group averaging. The data of control and experimental groups were normalized for maximal activity of a particular muscle within the gait cycle of the control group.

Averaged peak knee flexion angles during loading response and the averaged peak values of hip and knee moments, generated by weakened proximal muscle groups, in various sub-phases of stance were statistically examined with an independent *t*-test.

3. Results

3.1. Muscle strength

Table 1 shows the results of isokinetic muscle strength assessment. The ratios between the mean values indicate,

Table 1
Results of isokinetic muscle strength assessment for both groups

	SMA group (Nm)	CONTROL group (Nm)	Ratio (%)
Dorsiflexors concentric	0.16 (0.08)	0.33 (0.05)	48
Dorsiflexors eccentric	0.51 (0.2)	0.77 (0.09)	66
Plantarflexors concentric	0.25 (0.22)	1.05 (0.26)	24
Plantarflexors eccentric	1.18 (0.51)	2.03 (0.54)	58
Knee extensors concentric	0.16 (0.12)	2.26 (0.52)	7
Knee extensors eccentric	0.33 (0.23)	3.45 (0.93)	9
Knee flexors concentric	0.17 (0.09)	1.09 (0.31)	15
Knee flexors eccentric	0.68 (0.35)	1.7 (0.42)	40

Given are mean values and standard deviations.

consistently with the clinical description, that proximal muscle groups in the SMA group were more impaired and extensors were more affected than flexors. Strength in the tested muscles was higher during eccentric exercise. The results showed significantly limited strength of knee extensors in the SMA group. Standard deviations were similar between the groups.

3.2. Temporal gait characteristics

The data on gait velocity, stride length and cadence are presented in Table 2. The SMA group exhibited shorter stride length, lower cadence and lower gait velocity. Even though clinically relevant these differences were not statistically significant.

3.3. Kinematics and kinetics

Kinematic patterns are shown in Fig. 1. Posterior trunk and anterior pelvic tilt were observed in the SMA group throughout the whole gait cycle. After heel strike a pronounced internal rotation of the pelvis characterized loading response and mid-stance in the SMA group. The internal rotation of the pelvis was associated with rapid hip extension during the first 25% of stance in the SMA group. The hip was more flexed and adducted throughout the majority of gait cycle. The sagittal knee joint graph showed no knee flexion during loading response, while the shape of the sagittal ankle joint graphs was similar in both groups and heel strike at initial contact was observed in both groups.

Increased hip adduction was accompanied by increased knee and ankle valgus.

Kinetic patterns are shown in Fig. 2. The hip flexion/extension moment profile showed smaller amplitude and shorter duration of hip extensor moment in the first half of stance phase. A smaller amplitude in hip flexor moment during the second half of stance phase was noted. In the first half of stance phase, absence of hip internal rotation moment was seen. This allowed for an associated movement of the pelvis into internal rotation after heel contact. This differed from normal walking, where the burst of internal hip rotation moment facilitated movement of the pelvis into external rotation. Furthermore, the normal short burst of hip adduction moment after initial contact was absent. The knee flexion/extension moment profile showed minimal extension moment generation throughout the whole gait cycle and particularly during the loading response (statistically significant, Table 2). Ankle plantarflexion/dorsiflexion moments and power profiles showed similar shapes in both groups. There was no knee power absorption/generation throughout the majority of stance phase in the SMA group, while the hip power was similar in both groups. Ground reaction forces showed similar shapes in the anterior/posterior and medio/lateral directions. The vertical component showed distinctive difference during loading response where the body weight transfer onto the landing limb was delayed. This was associated with prolonged activity of the contralateral hip abductors, as suggested by the prolonged hip abductor moment at the end of the stance phase.

Table 2
Temporal gait characteristics and selected kinematic and kinetic values for both groups

	CONTROL (<i>n</i> = 9)	SMA (<i>n</i> = 7)	<i>p</i> -Value (<i>t</i> -test)
Cadence (steps/min)	107.61 (8.06)	94.3 (9.54)	0.815
Gait velocity (m/s)	1.22 (0.18)	0.84 (0.10)	0.146
Stride length (m)	1.37 (0.14)	1.08 (0.16)	0.389
Peak knee flexion–loading response (°)	15.09 (5.19)	3.91 (4.58)	0.001*
Peak knee extension moment–loading response (Nm/kg)	0.37 (0.20)	0.04 (0.06)	0.001*
Peak hip extension moment–loading response (Nm/kg)	0.51 (0.21)	0.28 (0.13)	0.027*
Peak hip flexion moment–pre-swing (Nm/kg)	−0.84 (0.08)	−0.55 (0.20)	0.001*
Peak hip adduction moment–loading response (Nm/kg)	−0.10 (0.12)	0.05 (0.07)	0.015*
Peak hip rotation moment–loading response (Nm/kg)	−0.10 (0.04)	−0.03 (0.02)	0.001*

Given are mean values and standard deviations.

* *p* < 0.05 considered as statistically significant.

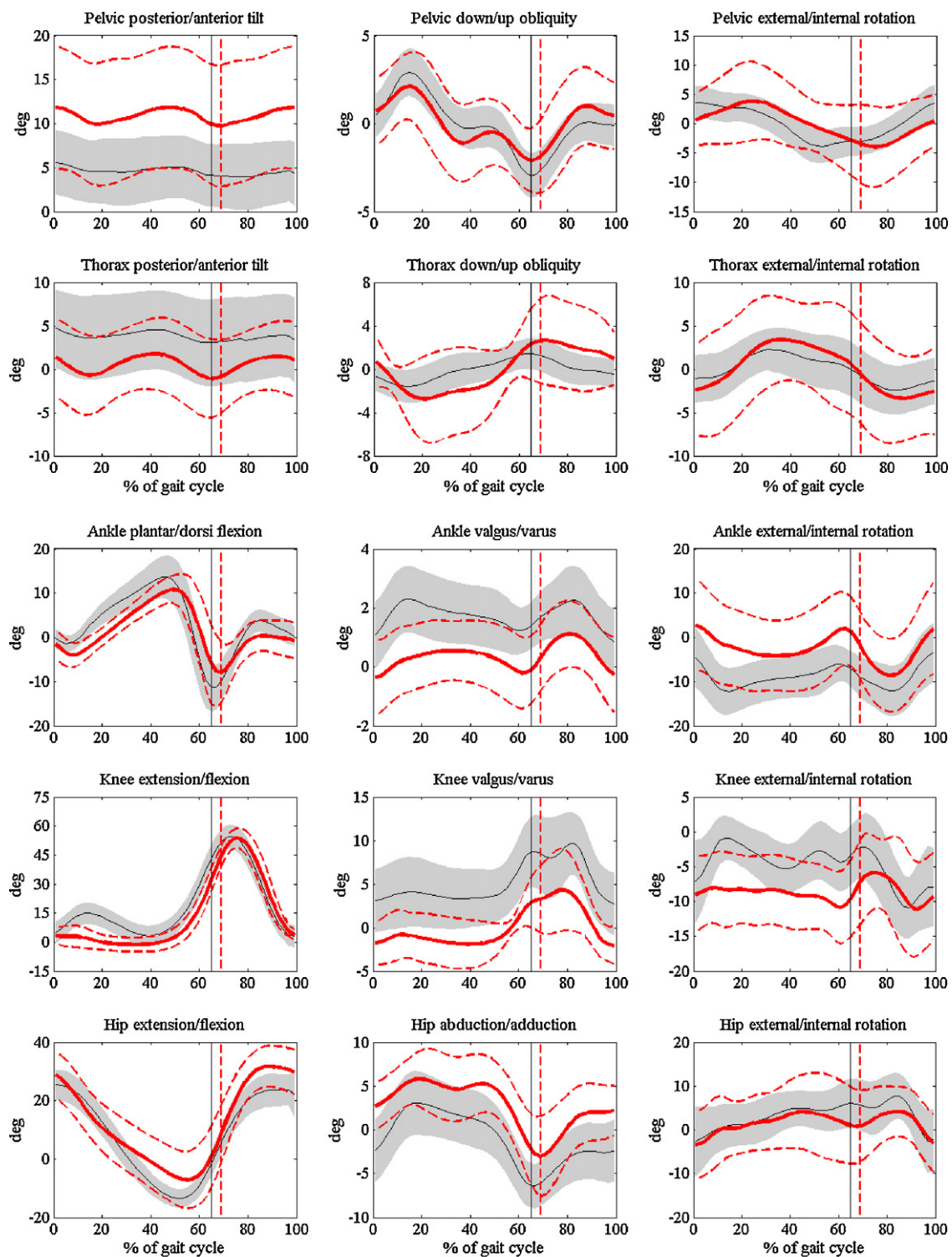


Fig. 1. Kinematic gait patterns showing averaged values and one standard deviation for SMA group (thick line: mean values and dotted lines: standard deviations) and CONTROL group (thin line: mean values, and shaded region: standard deviations). Negative values on ordinal graphs' axes correspond to firstly stated and positive values correspond to secondly stated movement direction in the graphs' titles. Toe-off is indicated by a vertical thin line (CONTROL) and vertical dotted thick line (SMA).

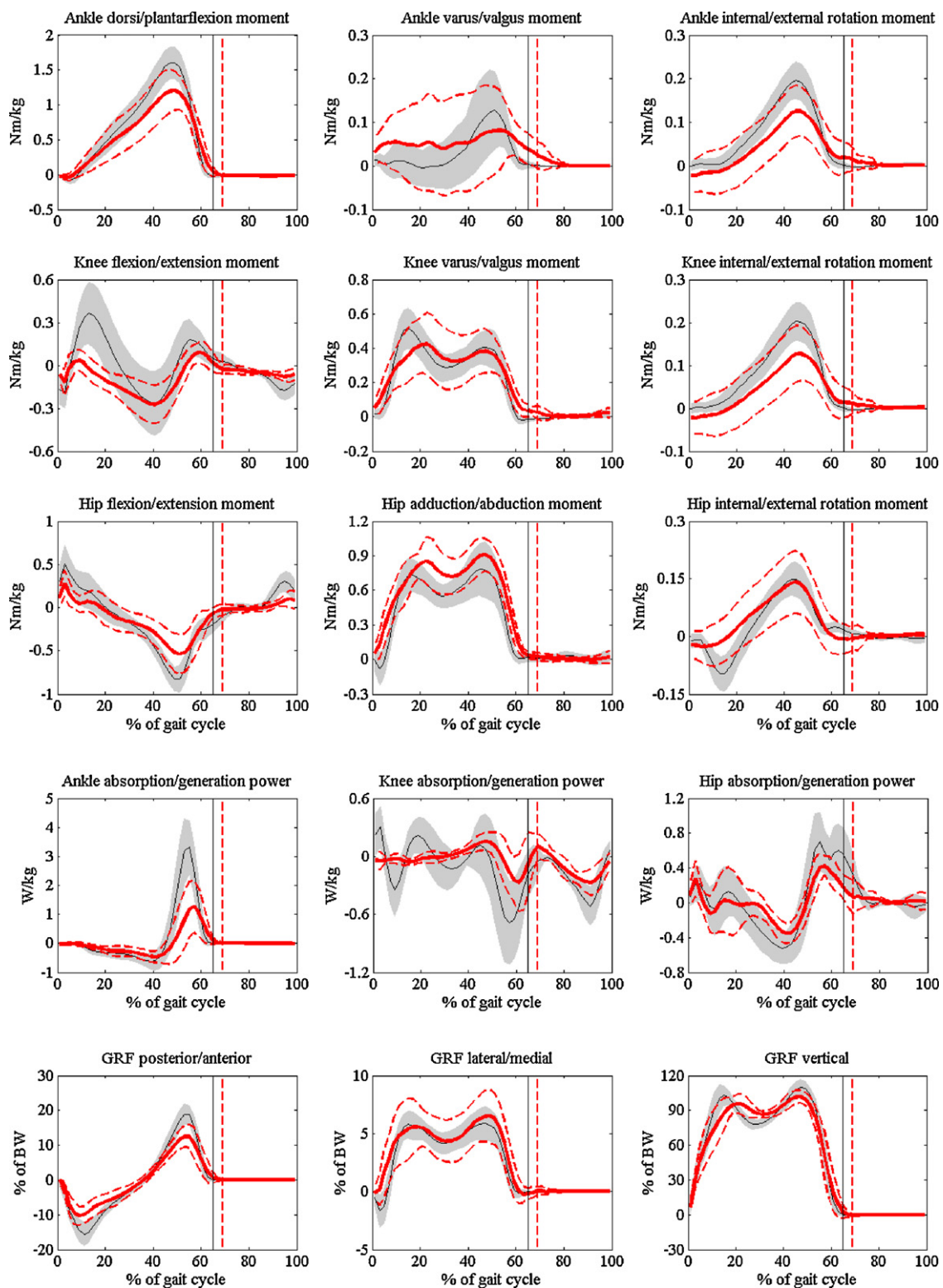


Fig. 2. Kinetic gait patterns showing averaged values and one standard deviation for the SMA group (thick line: mean values and dotted lines: standard deviations) and CONTROL group (thin line: mean values, and shaded region: standard deviations). Negative values on ordinal graphs' axes correspond to firstly stated and positive values correspond to secondly stated movement direction in the graphs' titles. Toe-off is indicated by a vertical thin line (CONTROL) and vertical dotted thick line (SMA).

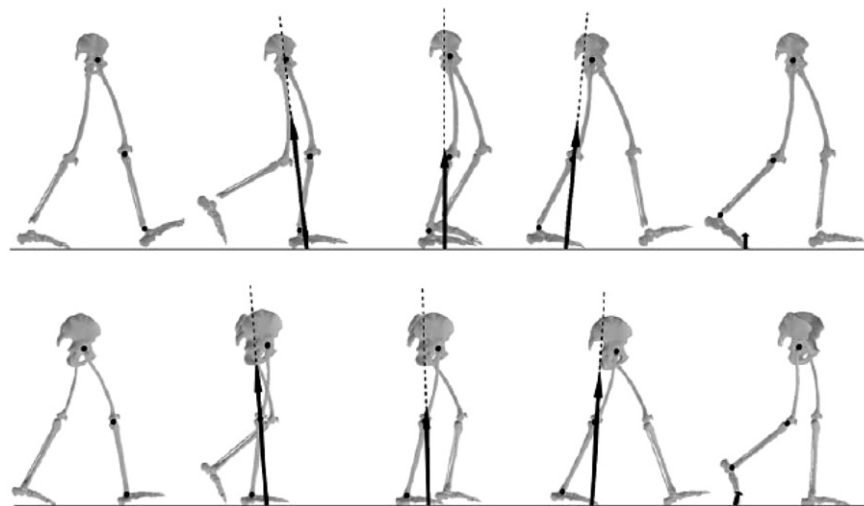


Fig. 3. Display of a lower body skeleton together with GRF and COP in five consecutive instants of stance phase that occurred approximately at 0%, 20%, 40%, 55% and 60% of a gait cycle. Upper panel: CONTROL and lower panel: SMA.

Fig. 3 illustrates kinematic and kinetic patterns of a CONTROL and an SMA subject. Visual comparison of five instants of stance phase revealed the differences in control of walking. Both subjects landed on the extended limb and on the

heel. At 20% of the stance phase the SMA subject had the pelvis tilted forward and rotated internally while the hip was considerably more extended as in the selected normal subject. The center of pressure (COP) was shifted more along the foot

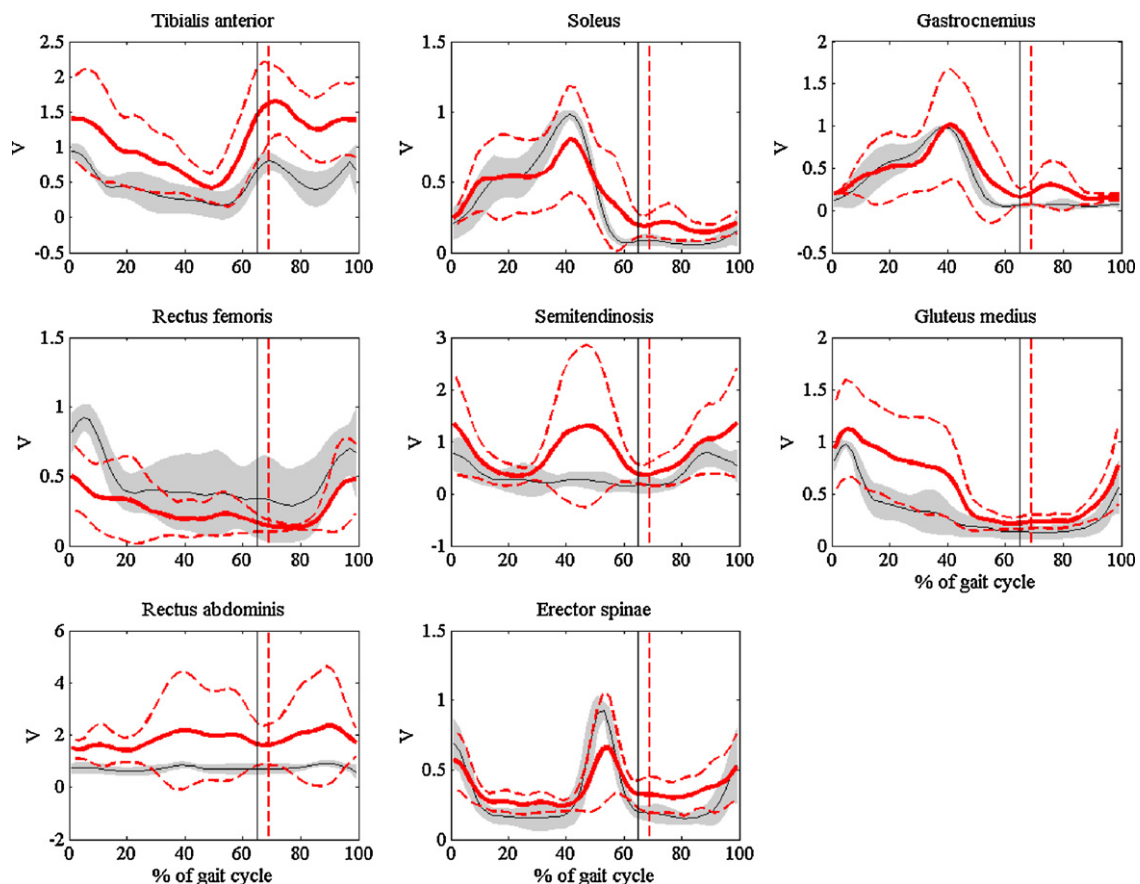


Fig. 4. EMG patterns showing averaged values and one standard deviation for SMA group (thick line: mean values, and dotted lines: standard deviations) and CONTROL group (thin line: mean values and shaded region: standard deviations). Toe-off is indicated by a vertical thin line (CONTROL) and vertical dotted thick line (SMA).

as compared to the normal subject. By assuming this posture the flexion moment on the knee was minimized while the ground reaction force (GRF) exerted an extension moment at the hip joint and the spine, leading to lumbar lordosis. Similar differences were also observed at 40% of the gait cycle. In the continuation of stance phase similar kinematic and kinetic patterns were seen in both subjects.

Fig. 4 shows EMG activity represented by mean envelopes and standard deviations. Visual comparison revealed premature activity in the soleus and gastrocnemius in the SMA group, which is associated with earlier displacement of COP along the foot (Fig. 3). Prolonged activity of gluteus medius was observed during loading response and mid-stance. This was probably associated with its role as a hip abductor, which would be supported by the associated prolonged duration of hip abductor moment. A distinct burst of activity was seen in semitendinosus during pre-swing associated with unlocking of the fully extended knee. Increased activity of rectus abdominis was noted throughout the gait cycle. Visual inspection of other muscles' EMG showed similarity in envelope shape between the groups.

4. Discussion

Muscle weakness limits walking ability in people with neuromuscular disorders. Coping responses include decrease of walking speed, step length and swing time in order to reduce mechanical output requirements of weakened muscles. One of the most debilitating combinations is simultaneous weakness of hip and knee extensors. One compensatory mechanism employed involves control of COP and GRF in such a way that appropriate external moments imposed by GRF onto the knee and hip joints are achieved. This was described by Sutherland et al. [5,6] for patients with DMD. The results of our study in SMA type III are in agreement with their findings. Furthermore, the results of our study identified a second compensation mechanism. This involves internal rotation of the pelvis in early stance, which facilitates rapid hip extension. This is further facilitated by the lack of hip internal rotation moment during the first 20% of stance. If this was present, it would drive the pelvis into external rotation in the transverse plane. With the above compensation mechanism, positioning GRF in front of the knee and behind the hip joint is further facilitated. This may explain increased internal rotation observed at the ankle, for which Sutherland et al. [5,6] did not find a plausible explanation. A third compensatory mechanism relates to decreased rate of weight acceptance immediately after foot contact. This can be seen from the time course of the vertical component of the ground reaction force. This is achieved by prolonged activity of hip abductors on the contralateral side. The EMG activity of gluteus medius during loading response and mid-stance, supports this explanation. The three described compensatory

mechanisms can effectively work in synergy only during double support stance phase where both legs are in contact with the ground, thus forming a closed kinematic chain. Overall, our results show that the patients with SMA type III adopt a control strategy, which minimizes external flexion moments produced by GRF on the knees, hips and spine.

When comparing our results with the results of Sutherland et al. [5,6], who investigated and described gait changes in three groups of DMD according to progression of disease, we can conclude that considerable similarity exists. Gait patterns were similar at comparable speeds and included excessive anterior pelvic tilt, lumbar lordosis, extended knee throughout the stance phase, internal pelvic rotation after foot contact facilitating rapid hip extension, increased hip flexion in swing allowing foot clearance and rapid displacement of COP after foot contact minimizing external moments on the knee, hip and trunk. The only difference between the two conditions lies in the foot contact, which is heel strike in SMA and footflat/forefoot in DMD. However, this is a result of more affected dorsiflexors in DMD [5], while functionally the set of compensatory changes allowing weight acceptance and progression remains the same. Armand et al. [8], who compared kinematics, kinetics and electromyographic recordings in a limited sample of SMA type II and DMD subjects, have suggested that different compensatory strategies may be employed by the two groups. However, the subjects they compared walked with different speed, step length and cadence.

Deymeer et al. [9] and Ueno et al. [11] have suggested that important differences exist between SMA and DMD in muscle involvement. Their findings support the currently accepted notion that in SMA muscle weakness around the hip is characterized by weak iliopsoas with relatively preserved gluteal muscles as opposed to DMD, where weak gluteal muscles with relatively preserved iliopsoas is seen. However, the results of our study show that these presumed differences in muscle involvement between the SMA and DMD [5] groups in the "transitional" stage of disease progression do not play a significant role and that kinematics and kinetics in both groups appear remarkably similar. The observed similarity in control of walking in SMA type III, could be attributed to the diminished strength of abdominal muscles (as established on clinical examination), compensated by anterior pelvic tilt and lumbar lordosis. Similar posture is also required in DMD, due to weakness of hip extensors.

Our study provides data on walking patterns in a group of individuals with SMA that is homogeneous. This is ascertained by the muscle strength assessment on the isokinetic dynamometer where a comparable variability was seen in the SMA and CONTROL groups. Unfortunately, due to methodological constraints in isokinetic hip muscle strength measurements and the proximal weakness of the tested patients with SMA type III, we were not able objectively to quantify their hip flexors and extensors strength.

A limitation of our study involves the difference in walking speed, step length and cadence between the two examined groups. This could have some influence on the validity of our interpretation of results and identification of compensatory changes: walking at lower speed decreases the amplitude of characteristic peaks of kinematic and kinetic patterns. However, according to the literature, moderate changes of $\pm 20\%$ from natural cadence – which is around 105 for adults – results in only minor differences in the shapes and amplitudes of joint kinematics and kinetics motor patterns [12]. Therefore, the distinct qualitative and quantitative changes in shape of some of the kinematic and kinetic motor patterns observed in the SMA group could be confidently viewed as compensatory changes.

The variability of kinematic and kinetic patterns was similar between groups. Therefore, the results of our study can also serve as a model of changed neural control of walking, caused by muscle weakness of proximal muscles of the lower limbs. Our results are in agreement with findings of the simulation study by Goldberg and Neptune [13] in which compensatory strategies in response to muscle weakness were examined. They concluded that compensatory activity of ankle plantarflexors is of critical importance when hip and knee extensors are weakened. In addition, our study has shown that compensatory activity of hip rotators and abductors represents an important contribution that works in synergy with compensatory ankle plantarflexors activity. Our findings have direct application to rehabilitation and/or preventive exercise programs which should both target maintenance of ankle plantarflexors strength and focus on maintaining strength of the hip rotators and abductors.

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Conflict of interest

None.

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