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Gait propulsion in patients with facioscapulohumeral muscular dystrophy and ankle plantarflexor weakness

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ABSTRACT

Facioscapulohumeral muscular dystrophy is a slowly progressive hereditary disorder resulting in fatty infiltration of eventually most skeletal muscles. Weakness of trunk and leg muscles causes problems with postural balance and gait, and is associated with an increased fall risk. Although drop foot and related tripping are common problems in FSHD, gait impairments are poorly documented. The effect of ankle plantarflexor involvement on gait propulsion has never been addressed. In addition to ankle plantarflexion, gait propulsion is generated through hip flexion and hip extension. Compensatory shifts between these propulsion sources occur when specific muscles are affected. Such a shift may be expected in patients with FSHD since the calves may show early fatty infiltration, whereas iliopsoas and gluteus maximus muscles are often spared for a longer time. In the current study, magnetic resonance imaging was used to assess the percentage of unaffected calf, iliopsoas and gluteus maximus muscles. Joint powers were analyzed in 10 patients with FSHD at comfortable and maximum walking speed to determine the contribution of ankle plantarflexor, hip flexor and hip extensor power to propulsion. Associations between muscle morphology, power generation and gait speed were assessed. Based on multivariate regression analysis, ankle plantarflexor power was the only factor that uniquely contributed to the explained variance of comfortable (R^2 = 80%) and maximum (R^2 = 86%) walking speed. Although the iliopsoas muscles were largely unaffected, they appeared to be sub-maximally recruited. This submaximal recruitment may be related to poor trunk stability, resulting in a disproportionate effect of calf muscle affliction on gait speed in patients with FSHD.

1. Introduction

Facioscapulohumeral muscular dystrophy (FSHD) is one of the most common hereditary neuromuscular disorders with a prevalence of 1:15,000–1:20,000 persons [1,2]. Although weakness typically starts in the muscles of the face and shoulder girdle, FSHD is characterized by progressive fat infiltration of eventually almost all skeletal muscles. Weakness of the trunk and lower extremity muscles, which is present in more than half of the patients [3], causes problems with postural balance and gait and is related to an increased risk of recurrent falls [4–8]. A typical gait impairment in FSHD is caused by early weakness of ankle dorsiflexors [6,9], leading to foot drop and an increased risk of tripping. This is often compensated by exaggerated hip and knee flexion to prevent tripping-related falls. However, recently, we were able to show that ankle plantarflexors are also involved in 56% of the people with FSHD [3], yet gait impairments due to ankle plantarflexor weakness in FSHD have been not yet been studied. Plantarflexor weakness may not only cause difficulties with the propulsion of gait, but has also been associated with an increased fall risk in healthy elderly [10,11]. The latter finding was related to a reduced capacity to recover from a trip due to a lack of compensatory push-off power.

It has been reported that comfortable walking speed in FSHD is on average 20% reduced compared to healthy controls [4,6,7], even in persons without clinically detectable lower extremity weakness [6]. Although walking speed was associated with hip flexion, knee extension and ankle dorsiflexion strength assessed by manual muscle testing [4], only one study has investigated gait kinetics in people with FSHD [6]. This study revealed that ankle kinetics was

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different from controls, which was related to typical drop-foot problems [6]. During normal walking, propulsion can be attributed to concentric activity of the ankle plantarflexors, hip flexors and hip extensors. As a consequence, the calf, iliopsoas and gluteus maximus muscles together constitute the main determinants of walking speed in healthy persons, of which the calf muscles are known to contribute most [12–15]. Previous studies have described a shift in the contribution to walking speed from ankle plantarflexor to hip flexor power in the case of impaired function or pain in the lower legs [16,17]. We hypothesize that people with FSHD and calf muscle weakness also tend to rely on such a trade-off phenomenon to maintain their preferred walking speed, since particularly the iliopsoas muscle appears to be preserved even in patients with a very long disease duration [3]. If so, people with FSHD would be able to compensate the functional consequences of calf muscle weakness for a relatively long time.

Hence, the aim of the current study was to examine the associations of walking speed with ankle plantarflexor, hip flexor and hip extensor power and relate these to the degree of fatty infiltration of the calf, iliopsoas and gluteus maximus muscles in FSHD.

Table 1
Ricci Clinical Severity Scale.

0.5	Facial weakness
1	Mild scapular involvement without limitation of arm abduction; no awareness of disease symptoms is possible
1.5	Moderate involvement of scapular and arm muscles or both (arm abduction MRC $>60^{\circ}$ and strength MRC ≥ 3 in arm muscles); no involvement of pelvic and leg muscles
2	Severe scapular involvement (arm abduction $<60^{\circ}$ on at least one side); strength MRC <3 in at least one muscular district of the arms; no involvement of pelvic and leg muscles
2.5	Tibioperoneal weakness; no weakness of pelvic and proximal leg muscles
3	Mild weakness of pelvic and proximal leg muscles or both (strength MRC ≥ 4 in all these muscles); able to stand up from a chair without support
3.5	Moderate weakness of pelvic and proximal leg muscles or both (strength MRC ≥ 3 in all these muscles); able to stand up from a chair with monolateral support
4	Severe weakness of pelvic and proximal leg muscles or both (strength MRC <3 in at least one of these muscles); able to stand up from a chair with double support; able to walk unaided
4.5	Unable to stand up from a chair; walking limited to several steps with support; may use wheelchair for most activities
5	Wheelchair bound

Muscle strength was evaluated by using the clinical Manual Muscle Testing Scale (Medical Research Council).

2. Methods

2.1. Participants

A sample of 10 adults with genetically confirmed FSHD type I or II was recruited via the rehabilitation and neurology departments of our university clinic. We purposely included a wide range of disease severities, from patients who did not yet subjectively experience lower extremity muscle weakness at the one end, to severely affected patients who were barely able to walk anymore on the other hand. Specific exclusion criteria were: the presence of other neurological diseases affecting ambulation, severe cardiopulmonary disease, metal implants (contra-indication for MRI) and pregnancy. During an intake visit a physiatrist checked the inclusion and exclusion criteria and determined the clinical severity score (CSS) as described by Ricci et al. [22] (Table 1). In addition, body weight and height were determined. At the same day the radiological assessments were performed. The gait assessments in the movement laboratory were done within 8 weeks after the intake. This study was approved by the local ethics committee. All subjects gave written informed consent.

2.2. Radiological assessments

The patients were examined with a 3.0 Tesla (T) MR system (Skyra; Siemens, Erlangen, Germany) using the body coil. Imaging was performed using transverse turbo spin echo T1 weighted sequences of the lower body and trunk. Ten transverse images were obtained in four different regions: the trunk at the level of vertebra L4, the pelvis between the symphysis and the anterior superior iliac spine, the upper leg at 15 cm proximal to the apex of the patella and the lower leg at 15 cm below the patella (TR 750, TE 9.4, turbo factor 3, matrix 256×256, slice thickness 5 mm, slice gap 1 mm).

Software of Siemens (InveonTM Research Workplace 4.0) was used for quantification of muscle involvement of 36 muscles within the scanned regions. For each muscle, we identified the transversal slice that captured the largest cross-sectional area and manually traced the original edges of the muscles. Within these edges, the amount of remaining muscle tissue was obtained by calculating the surface area of a region of interest with a window width of 200 Hounsfield units which excluded both edema and fat tissue. This window level was adapted according to the brightness of the image. The proportion of remaining muscle tissue (PMT) was calculated as the cross-sectional area of the remaining muscle tissue divided by the cross-sectional area of the original muscle edges. Asymmetry values were calculated as the absolute difference in PMT-value between the left and right body side.

2.3. Gait assessments

Subject walked barefoot wearing shorts and a sleeveless top. Reflective markers were placed on the skin according to the PlugInGait full-body model (BodyBuilder, Vicon Motion Systems, Lake Forest, CA). Kinematic data were acquired using a six-camera Vicon motion analysis system (Vicon MX, Oxford Metrics, Oxford, UK) with a sample frequency of 100 Hz. Two AMTI (Advanced Medical Technology Inc., Watertown, MA, USA) force plates, embedded in a 10 m walkway, were used to record ground reaction forces at a sample frequency of 2400 Hz.

The subjects were instructed to walk over the walkway at their comfortable (V_{comf}) and at their maximum (V_{max}) walking speed. Once subjects were accustomed to walking on the walkway, three trials of both walking speeds were recorded, starting with comfortable walking. No information was given regarding the presence or position of the force plates to prevent any adaptations of step length or walking speed to their location.

Marker data was filtered with a low-pass fourth-order Butterworth filter with a cut-off frequency of 6 Hz and processed with the Vicon Clinical Manager model (VCM) to calculate full body kinematics and joint moments. Force plate data was filtered with a fourth-order Butterworth filter with a cut-off frequency of 6 Hz. Gait cycle events were detected based on force plate and marker position data.

Walking speed was calculated by dividing the traveled distance of the center of mass during two strides by the time passed in this timeframe. The mean walking speed was calculated by taking the average of three trials for both comfortable and maximum velocity. Ankle and hip powers were calculated as the product of the joint moment and the joint angular velocity. Peak hip extensor power (H1), generated mainly by the gluteus maximus, was determined during early stance. Peak ankle plantarflexor power (A2), generated by the calf, and hip flexor power (H3), generated mainly by the