BALANCE CONTROL IN PATIENTS WITH DISTAL VERSUS PROXIMAL MUSCLE WEAKNESS


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Abstract—Muscle weakness is consistently associated with falls in the elderly people, typically when present along with other risk factors. However, it remains unknown whether and how muscle weakness alone affects balance. This hampers development of more effective fall prevention strategies. Clinical observations suggest that the amount and distribution of muscle weakness influences balance control. We therefore investigated balance corrections in patients with either predominantly proximal (limb girdle muscular dystrophy (LGMD); n=8) or distal (distal spinal muscular atrophy; n=8) leg weakness, and 27 matched healthy controls. Balance was perturbed using surface tilt rotations that were delivered randomly in eight directions. Balance measures were full body kinematics and surface electromyographic activity (EMG) of leg, arm, and trunk muscles. Both patient groups were more unstable than controls, as reflected by greater excursions of the centre of mass (COM), especially in the pitch (anterior–posterior (AP)) plane. COM displacements were greater in distal weakness patients. Patients with distal weakness had excessive and unstable trunk, knee and ankle movements, and this was present following both forward and backward directed balance perturbations, possibly reflecting the greater use of distal leg muscles in these directions. In contrast, the less weak proximal weakness patients demonstrated unstable trunk and ankle movements only for backward directed balance perturbations, possibly reflecting the greater use of distal leg muscles in these directions. In contrast, the less weak proximal weakness patients demonstrated unstable trunk and ankle movements only for backward directed balance perturbations. Both patient groups used arm movements to compensate for their instability. We conclude that primarily distal but also proximal muscle weakness leads to significant postural instability. This observation, together with the retained ability of patients to use compensatory arm movements, provides targets that may be amenable to improvement with therapeutic intervention.

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Key words: balance control, dynamic posturography, limb girdle muscular dystrophy, distal spinal muscular atrophy. Adequate balance control depends on the presence of reliable “afferent” information, generated by visual, vestibular and proprioceptive sensory inputs. This afferent information needs to be centrally processed and integrated, before it is translated into effective postural corrections once a balance perturbation has occurred. This “afferent” component of the balance repertoire has multiple components, involving both the CNS (e.g. the pyramidal tract) and the peripheral nervous system (ranging from motor neurons to muscles). It is becoming increasingly clear that adequate muscle strength is a key element for stable postural control. Indeed, epidemiological surveys have consistently identified muscle weakness as an important risk factor for falls in the elderly people (Moreland et al., 2004; Horlings et al., 2008).

However, weakness in the elderly people is typically present along with multiple other risk factors, leaving it uncertain whether muscle weakness alone affects balance control. Evidence that this may be true stems from observations of falls and postural instability in patients suffering from neuromuscular disorders such as facioscapulo–humeral muscular dystrophy, polymyositis or myotonic dystrophy (Lord et al., 2002; Pieterse et al., 2006; Wiles et al., 2006). However, very little is known about the pathophysiology of instability and falls in such patient populations. Moreover, even less is known about how muscle weakness affects postural stability and leads to falls (Horlings et al., 2008). Indeed, only very few studies have studied balance control in subjects with muscle weakness (Barrett et al., 1988; Lord et al., 2002; Lehmann et al., 2006), and no prior study has used advanced posturography techniques to analyse balance corrections in detail. This lack of knowledge currently hampers development of more effective fall prevention strategies.

We therefore decided to test balance reactions in patients with isolated muscle weakness. We were specifically interested whether the distribution of muscle weakness would be important, for two reasons. First, anecdotal clinical observations suggested that patients with distal leg weakness are particularly prone to stumbling, while patients with more proximal leg weakness would be more unstable following external balance perturbations (Horlings et al., 2008). Second, prior studies using dynamic posturography provided a theoretical framework to suggest that balance correcting synergies could be specifically altered, depending on the site of muscle weakness. It has been shown that some muscle responses are particularly sensitive to balance perturbations in the pitch (anterior–posterior (AP)) plane, while others are more sensitive to balance perturbations in the roll (side-to-side) plane or a...
combination of these two planes (Henry et al., 1998; Carpenter et al., 1999; Küng et al., 2009a). And these muscle response sensitivities appear to follow a proximal-to-distal distribution, proximal and axial muscles (such as paraspinals or gluteus medius) being more roll-oriented (Carpenter et al., 1999), lower leg muscles being more pitch-oriented and knee muscles acting in both directions (Küng et al., 2009b).

Based on these directional sensitivities, the question arises whether patients suffering from weakness in distal leg muscles have more difficulty maintaining balance in the pitch plane, and those suffering from proximal leg muscle weakness in the roll plane. Such knowledge can also help to fine-tune therapeutic interventions. For example, if patients with pure proximal weakness truly have unstable roll-directed balance correcting strategies, they would require a different intervention than those suffering from pure distal weakness with presumably unstable pitch-directed strategies.

These issues can be addressed by examining patients with specific weakness patterns. For example, limb girdle muscular dystrophy (LGMD) is predominantly characterised by pure proximal weakness of the trunk, arms and legs, and could serve to investigate the pathophysiology of instability caused by proximal weakness. Conversely, distal spinal muscular atrophy (SMA) patients experience pure distal weakness of the legs and arms, which does not progress proximally, thus providing pathophysiological insights into the effect of distal weakness on instability. However, it should be borne in mind that even if these patients have no additional clinical symptoms, such as sensory deficits, that could influence balance, they generally have population differences in extent of disabling muscle weakness.

Our goal was to examine the specific patterns of instability seen in patients with either proximal or distal muscle weakness. To analyse balance corrections in detail, we used a multi-directional rotating support surface with multimodal outcome measures (EMG (electromyographic), kinematics and kinematics). We hypothesised that distal muscle weakness would cause instability in the pitch plane, and that proximal muscle weakness would cause instability in the roll plane. Furthermore, we expected each group to employ different movement strategies—based on adapted muscle synergies—to maintain upright balance.

**EXPERIMENTAL PROCEDURES**

**Subjects**

We included eight patients with proximal weakness caused by LGMD (PROXIMAL), five patients with distal weakness caused by distal SMA (DISTAL), and 27 healthy age- and sex matched controls. All, but one, PROXIMAL patients had LGMD type 1B, which is characterised by pure proximal muscle weakness without sensory deficits. One patient showed a very similar muscle weakness pattern, but the LGMD had not been specified in this patient yet. Patients were recruited after a careful selection procedure from the large outpatient population at the Radboud University Nijmegen medical centre, The Netherlands. All patients were thoroughly screened and tested to have pure proximal or pure distal

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General characteristics for the distal weakness, proximal weakness, and control subjects and the mean population results and standard error of the mean (SEM) of the manual muscle testing for the patients. The means of right and left muscles are given. P-values for 3-group analysis of variance (ANOVA) (first four rows) or nonparametric Mann–Whitney (remaining 17 rows) testing are given. ns, not significant (P>0.05).

Leg weakness without any sensory deficits in the lower extremities that might influence balance. This included assessment of vibration sense (with Rydell-Seiffer tuning fork [Martina et al., 1998]), pain sense, and touch sense in legs, both proximally and distally; movement sense was tested in distal legs. Achilles tendon and patellar tendon reflexes were also tested to be normal. Patients with other neurological or orthopaedic disorders were also excluded, as were patients using medication that might influence balance. Patients had to be able to stand and walk independently. Patients and controls did not differ with respect to age, height, and weight and body mass index (Table 1). Subjects completed a brief questionnaire concerning mobility and balance in daily life, including the activity based confidence scale (ABC). Muscle strength was assessed in all patients according to the manual muscle testing medical research council (MRC)-scale adapted from Kendall (1993). This adapted scale allows scoring muscle strength from zero (no contraction) to 10 (normal strength). Posturographic testing took place at the University Hospital, Basel, Switzerland. All subjects gave witnessed written informed consent to participate in the experiments according to the declaration of Helsinki. The institutional ethical review boards of the University Hospital of Basel and Radboud University Nijmegen medical centre approved the study. To minimize subjects from suffering, two persons stood next to platform to avoid the subject from falling and hand rails were available.

**Procedure and measurement system**

Subjects stood on a servo-controlled platform that could tilt in combinations of the pitch and roll directions. The roll and the pitch axis of the platform had the same height which was equal to the average distance of the ankle joint to the soles of the feet. The subjects’ feet were tightly strapped into heel guides which were
fixed to the upper surface of the movable platform and prevented stepping reactions when stance perturbations occurred. The roll axis passed between the feet. The stance width was standardised (14 cm) and two handrails of adjustable height were located 40 cm from the sides of the platform centre. Subjects were informed that they were allowed to grasp the handrails if they needed support. Two assistants were present to lend support in case of a fall. The test protocol was identical to that of Bakker et al. (2006). In summary, stimuli consisted of rotations of the platform in eight different directions with a constant velocity of 60 deg/s and a constant amplitude of 7.5 degrees. Perturbation directions were pure forwards (toes down, 0°), forward right (45°), pure right (90°), backwards right (135°), pure backwards (toes up, 180°), backwards left (225°), pure left (270°), and forward left (335°). Each perturbation direction was presented randomly ten times. The first trial was excluded from the analysis to avoid 1st trial and habituation effects entering the data.

Data collection

Recordings of biomechanical and EMG data commenced 100 ms prior to perturbation onset and were collected for a total of 1 s. To record EMG activity, pairs of silver–silver chloride electrodes were placed unilaterally approximately 3 cm apart along the muscle bellies of left tibialis anterior, left soleus, left biceps femoris (hamstrings), left rectus femoris (quadriceps), left gluteus medius, left deltoid medius (ganz acromialis) and bilaterally on lower (L1–L2) and upper (Th4–5) paraspinals muscles. EMG recordings were analogue band-pass filtered between 60 and 600 Hz, full-wave rectified and low-pass filtered at 100 Hz prior to sampling at 1 kHz.

Full body kinematic data were collected using a three-dimensional optical tracking system with 21 infrared emitting diodes (IREDs) (Optotrak, Northern Digital). The optotrak cameras sampled the IRED signals at 64 Hz and were placed approximately 4 m in front of the subject. IREDs were placed bilaterally on the ankle, knee, greater trochanter, anterior superior iliac spine, wrist, elbow, shoulder, above the ears, and on the angulus sternum and the chin. Three IREDs were placed on the rotating platform to define the pitch and roll movements of the support surface during perturbation in addition to potentiometer outputs on the axes. Subjects wore tight-fitting shorts and vests to prevent marker movements with respect to the skin.

Support surface reaction forces of both feet were measured from strain gauges embedded within the rotating support surface. The strain gauges were located under the corners of the plate supporting each foot. From forces recorded perpendicular to the support-surface by the strain gauges under the left/right foot and the distances to the centre of ankle joint rotation, the AP and lateral ankle torques were calculated for the left/right foot. The torques from the left and right foot were added together and displayed to the subject as excursions on two rows of diodes mounted on a cross 4 m from the subject, in order that the subject could control his/her pre-stimulus position.

Data analysis

Primary variables of interest were centre of mass (COM) displacement and velocity, trunk, pelvis, knee, and ankle angle and angular velocity profiles as well as muscle EMG responses of the leg, arm and trunk. Following analogue to digital data conversion of EMG signals, these were averaged off-line with Optotrak® across each perturbation direction. Zero latency was defined as the onset of platform rotation. Subject averages were pooled to produce sample population averages for a single direction. Biomechanical Optotrak® data were digitally filtered at 16 Hz using a zero phase shift 4th order Butterworth filter. Total body COM displacement was calculated separately for the anterior-posterior (AP), medio-lateral (ML) and vertical directions using a 12 segment adaptation (see Visser et al., 2008) of a 14 segment model of the body (Winter et al., 2003). Two trunk segments (upper and lower trunk) were used instead of four. Very occasionally the DISTAL patients (not always the same subject) raised their arms considerably to prevent falling, causing IRED signals at the wrists to be lost from view by the Optotrak® system due to accompanying arm supination. Therefore we calculated the COM position and velocity excluding wrist markers in the model. That is the body was modelled without the arms.

We calculated the following angular displacements: absolute roll and pitch trunk angle, absolute roll and pitch pelvis angle, ankle joint, knee joint, and shoulder–joint angles. Absolute rotation angles of the planes defined by trunk and pelvis body segments and the platform surface were defined using three or four markers on these segments. Knee and ankle joint angles were calculated using angles between segments either side of the joint (Bakker et al., 2006). Stimulus induced changes were calculated with respect to a pre-trigger time interval of 90 ms ending 10 ms prior to stimulus onset.

Each EMG response was corrected for background activity by subtracting the average level of pre-stimulus activity measured over a 90 ms period ending 10 ms prior to perturbation onset. The onset of activity was defined for each muscle based on the direction showing the greatest peak activity. From peak activity, the analysis algorithm looked backwards in time until activity first reached lower than mean pre-stimulus activity plus 2.5 σ. Then areas were calculated over 150 ms from this onset time for each individual response. A second response area was calculated over the next 200 ms after the first interval and a third over a fixed interval from 500 to 800 ms post stimulus with similar calculations used by Bakker et al. (2006).

Statistical analysis

Anthropometric differences between the groups were evaluated using analysis of variance (ANOVA) and post hoc testing (Bonferroni). For each dependent variable of interest a two-way repeated measures ANOVA was performed with group as between and direction as within factor using a significance level of 0.05. Significant differences between groups were evaluated within perturbation direction using Bonferroni post hoc tests.

RESULTS

Clinical details

DISTAL patients had less balance confidence during daily activities than PROXIMAL patients, as reflected by lower scores on the ABC scale (Table 1). DISTAL patients also reported a higher frequency of falls; three out of five DISTAL patients (60%) had fallen in the preceding month, and all DISTAL patients reported at least one prior fall in the past year. In contrast, only four of the eight PROXIMAL patients (50%) reported falls in the past year. All patients with falls also reported injuries, mainly bruises and scratches. Results of manual muscle testing in both patient groups are also reported in Table 1. PROXIMAL patients had significantly weaker upper leg muscles, and DISTAL patients had weaker lower leg muscles. All controls had normal strength. Fig. 1a visualises the mean muscle weakness present in PROXIMAL and DISTAL patients (Pieterse et al., 2007).

COM displacement

Fig. 1b shows stick-figures for a control subject, a DISTAL patient and a PROXIMAL patient while being perturbed backwards. These stick-figures display body segment motion measured by the infra-red sensors during the first
second directly after the perturbation has occurred. The control subject was stable, but both patients showed clear instability, and more so in the DISTAL patient compared to the PROXIMAL patient.

Fig. 2a shows the movements of the COM for the three populations, and for three perturbation directions (purely forward (toes-down), purely backward (toes-up), and purely sideways (rightward)). DISTAL patients showed greatest instability of the COM, especially while being perturbed backward (toes-up, 180°). COM velocity traces are depicted in Fig. 2b, showing greatest velocities for DISTAL patients, some 400 ms after perturbation onset, i.e. following onset of balance correcting muscle response. Early biomechanical responses to the perturbations (0–200 ms after perturbation onset) were similar for all three populations. As expected from the COM position traces in Fig. 2a, the backwards perturbation induced greatest COM velocity in both patient populations (Figs. 2b, 3a).

Fig. 3a shows a polar plot comparing the COM velocity in the three populations at the time of maximum velocity peak of the DISTAL patients (as indicated in Fig. 2b) for each perturbation direction. DISTAL patients showed greatest velocities, especially in the pitch directions (Fig. 3a). Significant differences for AP COM velocity between the groups were apparent for all directions (P<0.05) except 45°. PROXIMAL patients showed greater COM AP

Fig. 1. Individual examples. Visualization of the mean muscle weakness existing in PROXIMAL and DISTAL patients is depicted in Figure (a) (Pieterse et al., 2007). See Table 1 for exact mean manual muscle testing (MMT) scores. Figure (b) shows stick figures representing body motion in the pitch (anterior–posterior (AP)) plane of a healthy subject, a PROXIMAL, and a DISTAL patient in response to a backwards perturbation (180°). Dark lines represent starting position of the body segments, lighter lines the position until 1 s after perturbation. For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.
velocity compared to the control group only for backwards perturbations (perturbation directions: 135°, 180°, and 225° in Fig. 3a). Lateral COM velocity was similar across all groups (data not shown). At 750–850 ms post-perturbation, a significantly greater COM AP displacement (P<0.05) occurred in DISTAL compared to PROXIMAL for all directions except 45°, 270°, and 315° (Fig. 3b). PROXIMAL patients had greater COM displacement compared to controls for the backward perturbations (135°, 180°, and 225° in Fig. 3b, P<0.05). Fig. 3c provides the equivalent COM pitch angle based on the AP displacements at 750–850 ms, to permit a comparison with joint and body segment angular motion, as described below.

Joint and body segment angular responses

In order to judge which segments gave rise to the changes in equivalent COM pitch angle shown in Fig. 3c, we examined angle changes at several body segments.

**Ankle and knee motion.** When being perturbed backwards, both DISTAL and PROXIMAL patients showed a change from imposed dorsiflexion to an increased destabilising ankle plantar flexion (Fig. 4a). When perturbed forwards a changeover from plantar flexion to increased ankle dorsiflexion occurred only in DISTAL patients and not PROXIMAL patients (Fig. 4b). Mean left ankle angle and significant differences for all perturbation directions are illustrated in Fig. 4c and provide correlates with the trends shown in Fig. 3c. Ankle angular velocity traces, especially at 400 ms post-perturbation, were increased for DISTAL patients compared to PROXIMAL and control subjects for both pitch perturbation directions, but only in PROXIMAL patients compared to controls for backwards perturbation directions (P<0.05, data not shown).

Particularly for forward directions, knee flexion was greater in DISTAL patients compared to PROXIMAL patients and controls (perturbation directions: 0°, 45° and 315° in Fig. 4d). In addition, DISTAL patients flexed the downhill knee when perturbed sideways (45°, 90°, and 135° in Fig. 4d). Mean velocity for both knees was increased in DISTAL patients for forwards perturbations (0°, 45°, and 315°, P<0.05) at 400 ms after perturbation, and returned to almost normal at 800 ms post-perturbation (data not shown).

**Pelvis and trunk motion.** Hardly any differences in pelvis motion were found between the groups. Pelvis pitch angular motion was normal for both patient groups; however, trunk pitch motion was in excess of the pitch motion of controls to backward perturbations (Fig. 5a). In contrast only that of DISTAL patients was forwards instead of backwards in response to forward tilts (Fig. 5b).

Mean trunk pitch angles and significant differences across groups are given in Fig. 5c. Trunk pitch flexion was
increased for DISTAL compared to controls across all directions, except 270°, and increased for PROXIMAL patients for backwards directions (135° and 180° in Fig. 5c). At 400 ms post-perturbation, trunk pitch velocity was also increased in PROXIMAL patients compared to controls for backwards perturbations only (135° and 180° in Fig. 5d). For DISTAL patients, trunk pitch velocity at 400 ms was greater for all perturbation directions except 90° (Fig. 5d). Trunk roll angle was increased for DISTAL for 135° at 800 ms. Trunk roll velocity was increased for DISTAL for 135° and 180° at 400 ms, and for 270° and 315° at 800 ms post-perturbation (data not shown).

**Arm motion.** Both DISTAL and PROXIMAL subjects used more arm motion than control subjects, presumably in an attempt to maintain balance. As both arms will affect balance control, the mean of left and right arm rotation and the difference in left and right arm abduction was calculated. The rotation action in DISTAL subjects is significantly more forwards (i.e. in the compensatory direction) for backwards perturbations (180°, 225°). However, in PROXIMAL patients, arm abduction is only significantly greater for leftwards perturbations (225°, 270° and 315°).

**Muscle responses**

Muscle response patterns were similar for DISTAL, PROXIMAL and control subjects. Differences in muscle response amplitudes were most obvious for DISTAL patients. When looking at the responses to forward perturbations (0°), for which ankle, knee and trunk angle changes were greater than controls, DISTAL patients showed increased upper paraspinal (Fig. 6a) and hamstring (Fig. 6b) muscle activity. This was particularly obvious at the late interval (500–800 ms, Fig. 6g, h). For backwards perturbations, DISTAL patients showed a tendency for reduced responses in
tibialis anterior (data not shown). At the late interval (500 – 800 ms after the perturbation), deltoid muscle responses were increased in DISTAL patients for all perturbation directions, except 270° \((P/H11021/0.05, \text{data not shown})\).

PROXIMAL patients showed a tendency of decreased hamstrings at peak amplitude (Fig. 6d, e) and lower paraspinal muscle activity (Fig. 6b), but only for backwards directed perturbations. PROXIMAL patients showed decreased quadriceps activity for forwards directed perturbations, especially at the late interval (Fig. 6c, f, i).

DISCUSSION

This study showed that both distal and proximal weakness caused instability after support surface perturbations. Both patient populations had more difficulty with pitch than roll directed perturbations possibly due to the greater involvement of lower and upper leg muscles in pitch control (Küng et al., 2009a). Distal weakness patients showed instability of the COM for forwards and backwards perturbations, whereas proximal weakness patients were mainly unstable for backwards perturbations. It should be noted, however, that the level of muscle weakness was greater in the distal patients.

It has been shown before that muscle strength is a consistent risk factor for falls (Horlings et al., 2008) and an independent predictor of postural sway in healthy older adults during standing on an unstable surface (Lord and Menz, 2000). In nursing home residents with a history of falling, knee and ankle strength (peak torque and power) is decreased, most prominent in ankle dorsiflexion (7.5 times less power), compared to those without a history of falling (Whipple et al., 1987). Knee extension strength is a signif-
significant determinant of static balance, accounting for 10% of the variance. It accounted for 26% of the variance in dynamic stability (timed figure eight) in women with osteoporosis (Carter et al., 2002). However, the question remained what the influence of distal versus proximal muscle weakness is on instability and balance control.

**Instability and balance correcting strategies in distal weakness patients**

Instability in the distal weakness patients appeared to result from the inability to resist body motion about the ankle joint when support surface tilt occurs. This is the primary mode of control for correcting balance perturbations in the pitch plane (Carpenter et al., 1999; Küng et al., 2009a; Termoz et al., 2008). For example, the tendency to fall backwards causes increased plantarflexion of the ankle joint when the support surface tilts backwards. The activation of tibialis anterior and quadriceps resists this ankle joint motion. Muscle responses of tibialis anterior and soleus in patients with distal weakness tended to be decreased, but were not significantly different from healthy controls. Neither were changes in the EMG patterns apparent. The inability to move the body forwards when being perturbed backwards, could be seen in increased ankle plantar flexion, whereas on forwards perturbations ankle dorsiflexion was increased as a result of the inability to move the body backwards. Compensation for increased forwards motion of the body was seen in increased hamstring activity, resulting in increased knee flexion for forwards perturbations. On backwards perturbations, flexing the trunk forwards aided stability, as did forwards rotation of the arms. Hamstrings activity was increased in patients with distal weakness not only for forwards but also for lateral perturbations for which the COM was remained stable. DISTAL patients were observed to have additional knee and trunk flexion after pitch directed surface pertur-
bations. This compensatory strategy was effective in the roll plane; in the pitch plane only partially so, as can be concluded from the obvious instability of the COM. Indeed, it has been shown that bilateral knee bending during a balance correction enhances instability (Oude Nijhuis et al., 2007), whereas unilateral bending of the uphill knee aids balance control (Küng et al., 2009b). A subject for future studies is whether training patients with distal leg weakness to use knee bending during balance correcting responses may help reduce their instability and thereby reduce the risk of falls.

**Instability and balance correcting strategies in proximal weakness patients**

Patients with proximal weakness showed greatest instability for backwards perturbations. In contrast to DISTAL patients, control of ankle and knee flexion for forward tilts in PROXIMAL patients was similar to that of controls and the trunk remained tilt backwards. However, for backwards perturbations both ankle and trunk motion was unstable as in DISTAL patients. Knee motion was normal across all perturbation directions in these patients, although we found reduced muscle strength in upper leg muscles. Manual muscle strength measures, however, often are the result of activity of several muscles simultaneously, and can therefore not be directly related to EMG measures, although a linear correlation exists for some muscles (Gubler-Hanna et al., 2007). Upper leg muscle response amplitudes displayed a tendency to be less than normal across all muscles for proximal weakness patients, but this trend was not significant. It must be noted, however, that a direct relationship between muscle activity amplitudes and joint torques was not explored for the patient groups of this study.

**Considerations**

The question arises what severity of weakness is the threshold for suffering instability and falls, and whether this threshold is different for distal and proximal muscle weakness. Pai and Patton (1997) have reported that an ankle dorsiflexor strength decrease of at least 51% of normal, plantar flexion strength decrease of at least 35% of normal, and a friction level (feet–floor) of 0.82 or smaller increased the risk of instability, when using an inverted pendulum.
deficits. Although it has been argued that muscle weakness in other muscles or sensory, vestibular or orthopaedic proximal muscle weakness, with no clinical signs of weakness alone for equally weak patients. Given the low numbers of muscle weakness patients available, such studies may be difficult to perform.

The question arises why lateral instability was not noted in the PROXIMAL patients. We investigated the instability occurring in the PROXIMAL patient with greatest muscle weakness. The results of this patient followed the population trend in the pitch plane, but not in the roll plane. In the roll plane, COM displacement was considerably worse than the mean. Thus, severe proximal muscle weakness leads to roll instability; severe distal weakness, however, does not. This suggests that the site of muscle weakness influences the type of instability and the severity the amount of instability. Further research comparing patients with similar site of weakness, but variation in severity are needed to determine the influence of weakness severity at a specific location on balance control. Another aspect that needs to be examined in detail is the effect on the site of weakness alone for equally weak patients. Given the low numbers of muscle weakness patients available, such studies may be difficult to perform.

Although the number of patients in our sample was small, the patients formed two very homogenic groups. All patients with proximal weakness, except one, had LGMD type 1 B, which is characterised by symmetrical proximal lower limb weakness, and all patients with distal weakness had distal SMA. We selected our patients on pure distal or proximal muscle weakness, with no clinical signs of weakness in other muscles or sensory, vestibular or orthopaedic deficits. Although it has been argued that muscle weakness impairs proprioceptive control of standing (Butler et al., 2008) we were not able to establish significant changes in stretch reflex responses in the muscles we recorded from.

CONCLUSION

In conclusion, distal muscle weakness causes instability primarily for pitch directed perturbations. Proximal muscle weakness causes a lesser instability, but further studies are needed to determine the effect of weakness severity on balance control. Compensating balance correcting strategies consisted of trunk and arm movements in both populations. Ankle and knee bending was particularly abnormal in distal weakness patients leading to instability. It is therefore an open question whether training the use of knee movements to aid balance responses in these patient populations, might reduce instability and prevent falls. Future studies should focus on formulating muscle weakness thresholds and correlating these with COM instability following tilt perturbations in order to assess whether an increased risk of falls exists.

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