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Falls and stumbles in myotonic dystrophy
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A fall may be defined as a loss of balance resulting in coming to rest on the floor or ground or on another object below knee level; a stumble can pragmatically be defined as losing balance but regaining it before a fall occurs. Falls potentially cause loss of confidence, restriction of activity, and injury but, in the elderly, multidisciplinary evaluation and measures may reduce falls. A third of subjects of 65 years or over have 1 fall per year, but there is little information on patients with muscle disease. Patients with myotonic dystrophy type 1 (DM1) may have reasons to be at higher risk of falls, including critical fixed weakness in lower limb muscle strength, gait speed, and 7-day ambulatory activity monitoring were recorded. Subjects returned a weekly card detailing stumbles and falls.

Methods: 13 sequential patients with DM1 from different kindreds were compared with 12 healthy volunteers. All subjects were evaluated using the Rivermead Mobility Index, Performance Oriented Mobility Assessment, and modified Activities Specific Balance Confidence scale. Measures of lower limb muscle strength, gait speed, and 7-day ambulatory activity monitoring were recorded. Subjects returned a weekly card detailing stumbles and falls.

Results: 11 of 13 patients (mean age 46.5 years, seven female) had 127 stumbles and 34 falls over the 13 weeks, compared with 10 of 12 healthy subjects (34.4 years, seven female) who had 26 stumbles and three falls. Patients were less active than healthy subjects but had more falls and stumbles per 5000 right steps taken (mean (SD) events, 0.21 (0.29) vs 0.02 (0.02), p = 0.007). Patients who fell (n = 6) had on average a lower Rivermead Mobility score, slower self-selected gait speed, and higher depression scores than those who did not.

Conclusions: DM1 patients stumble or fall about 10 times more often than healthy volunteers. Routine inquiry about falls and stumbles is justified. A study of multidisciplinary intervention to reduce the risk of falls seems warranted.

Data collection
The muscle clinic record was used to extract retrospective information about previous illnesses, cardiac disorders, blood pressure, dizziness, blackouts, epilepsy, ocular problems, other problems recognised to contribute to falls or loss of consciousness, and other factors relating to mobility—for example, alcohol intake and drug treatment.

Methods

Subjects
Thirteen successive patients with DM1 from different families were recruited from our muscle clinic. Inclusion criteria were: patients aged ≥18 years; genetically confirmed diagnosis of DM1; from separate households in or near Cardiff; and sufficient cognitive and behavioural function to provide informed consent and participate. Twelve healthy volunteers were recruited from the local community. Exclusion criteria were a known history of any (other) major locomotor or neuromuscular disorder recognised to cause falls.

The study received local research ethics committee and Hospital Trust R&D approval. All subjects gave written informed consent.

Abbreviations: ABC-UK, modified activities-specific balance confidence scale; DM1, myotonic dystrophy type 1; HADS, Hospital Depression and Anxiety Scale; POMA, Performance Oriented Mobility Assessment; RMI, Rivermead Mobility Index

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Patients and volunteers were similarly matched for sex, height, weight, and body mass index, but the volunteers were younger (table 1). Patients had significantly more comorbidities for cardiac disorder, eye problems and cataract, and complaints of impaired balance, and they had higher scores on the depression subscale of HADS. Eight patients and four volunteers reported stumbles (four patients and one volunteer, all on sedative drugs). One patient died in the later stages of the study.

Patients had a significantly slower self-selected walking speed, shorter stride length, and reduced cadence compared with healthy volunteers (table 1). Patients were more inactive, had lower mean daily step counts (average 5000 steps), and lower peak and sustained walking speed, compared with healthy volunteers (mean (SD) 26 stumbles and three falls). When falls and stumbles were expressed in relation to activity, patients had 0.206 (0.288) events per 5000 steps taken (mean (SD)), compared with 0.021 (0.021) in healthy volunteers (p = 0.007).

In volunteers falls were usually from extrinsic causes (for example, a slippery surface) in an unfamiliar outdoor environment, while in patients intrinsic causes were more likely (legs “giving out”, “unsteadiness”, misperception of the environment) (table 2). Stumbles followed a similar pattern although many could not be classified in patients. Two patients attended hospital following falls; neither was admitted or sustained a fracture. In weeks 14–26, following which most subjects (10 of 12 patients and 10 of 12 healthy volunteers) returned a single report card, two patients reported further falls, with one fracturing a finger; four patients and four volunteers reported stumbles.

Associations tested across all subjects indicated that, although self-confidence about falling (ABC) was correlated with predicted liability to fall (POMA), there was no correlation between these scores and the actual number of stumbles or falls. Ankle dorsiflexion and plantarflexion, knee extension, and hip abduction strength were inversely correlated overall with the rate of falls and stumbles per 5000 steps (Spearman r, p<0.01).

Within the patient group alone ankle plantarflexion and dorsiflexion strength had the highest correlations with gait speed (Spearman r = 0.92, p<0.001, n = 10, and r = 0.75,

| Table 1 Demographics, gait, mobility, and lower limb strength in the study populations |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| Variable                        | DM1 (n = 13; 7f) | Volunteers (n = 12; 7f) | p Value |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| Age (years)                     | 46.5 ± 1.68     | 34.4 ± 1.73     | 0.002           |
| Height (m)                      | 1.68 ± 0.08     | 1.73 ± 0.08     | 0.1             |
| Weight (kg)                     | 75.6 ± 16.2     | 77.4 ± 15.8     | 0.8             |
| Body mass index (kg/m²)         | 27.1 ± 6.7      | 25.7 ± 5.2      | 0.6             |
| Gait indices                    |                 |                 |                 |
| Speed (m/s)                     | 0.83 ± 0.36     | 1.47 ± 0.16     | <0.001          |
| Stride (m)                      | 0.98 ± 0.32     | 1.53 ± 0.15     | <0.001          |
| Cadence (steps/min)             | 97.3 ± 19.7     | 114.6 ± 8.08    | 0.01            |
| Activity indices                |                 |                 |                 |
| Mean daily step count           | 3445 ± 1967     | 6324 ± 1222     | <0.001          |
| Peak 30 min (steps/min)         | 32.7 ± 11.2     | 52.5 ± 4.7      | <0.001          |
| Sustained 60 min (steps/min)    | 13.0 ± 8.5      | 24.8 ± 8.0      | 0.002           |
| Time inactive (%)               | 80.5 ± 9.2      | 72.1 ± 4.9      | 0.01            |
| Strength (N)                    |                 |                 |                 |
| Ankle dorsiflexion              | 54.3 ± 28.5     | 180.6 ± 67.6    | <0.001          |
| Ankle plantarflexion           | 109.4 ± 65.8    | 313.1 ± 126.3   | <0.001          |
| Knee flexion                    | 95.6 ± 50.7     | 143.7 ± 63.6    | 0.06            |
| Knee extension                  | 169.5 ± 72.9    | 274.4 ± 130.0   | 0.03            |
| Hip extension                   | 141.0 ± 57.8    | 262.0 ± 145.0   | 0.02            |
| Hip abduction                   | 96.5 ± 53.8     | 194.8 ± 91.6    | 0.006           |
| Mobility scales                 |                 |                 |                 |
| RMI (0–15)                      | 11.3 ± 3.9      | 15.0 ± 0        | 0.002           |
| POMA (0–28)                     | 20.6 ± 8.4      | 28 ± 0          | 0.008           |
| ABC-UK (%)                      | 60.6 ± 32.6     | 98.8 ± 1.4      | <0.001          |

*p = 11 for strength and mobility scales data. +p = 12 for strength and mobility scales data. \*n = 11 for strength and mobility scales data. \*n = 12 for strength and mobility scales data. DDaily right step count averaged over seven days. ABC-UK, modified activities-specific balance confidence scale; DM1, myotonic dystrophy type 1; f, female; N, newtons; POMA, Performance Oriented Mobility Assessment; RMI, Rivermead Mobility Index.

During weekly data collection the number of patients and volunteers who reported stumbles was similar (table 2) but more patients had a fall (NS). Eleven patients had 161 events (127 stumbles and 34 falls), while 10 volunteers had 29 events (26 stumbles and three falls). When falls and stumbles were expressed in relation to activity, patients had 0.206 (0.288) events per 5000 steps taken (mean (SD)), compared with 0.021 (0.021) in healthy volunteers (p = 0.007).

In volunteers falls were usually from extrinsic causes (for example, a slippery surface) in an unfamiliar outdoor environment, while in patients intrinsic causes were more likely (legs “giving out”, “unsteadiness”, misperception of the environment) (table 2). Stumbles followed a similar pattern although many could not be classified in patients. Two patients attended hospital following falls; neither was admitted or sustained a fracture. In weeks 14–26, following which most subjects (10 of 12 patients and 10 of 12 healthy volunteers) returned a single report card, two patients reported further falls, with one fracturing a finger; four patients and four volunteers reported stumbles.

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Within the patient group alone ankle plantarflexion and dorsiflexion strength had the highest correlations with gait speed (Spearman r = 0.92, p<0.001, n = 10, and r = 0.75,
p < 0.008, n = 11, respectively) and mean daily step count
(r = 0.56, p = 0.09, n = 10, and r = 0.59, p = 0.06, n = 11).
Compared with seven non-falling DM1 patients, six who did
fall had lower mean (SD) RMI scores (8.3 (3.4) vs 13.9 (2.2);
p = 0.005), were more likely to use aids or a person to assist
mobility indoors (3/6 vs 0/7) or outdoors (5/6 vs 1/7) (Fisher's
exact test, p = 0.07; p = 0.08), had a slower self selected gait
speed (0.61 (0.38) vs 1.02 (0.24) m.s⁻¹; p = 0.05), and had
higher depression scores (8.7 (3.3) vs 3.7 (2.4); p = 0.01). A
backward regression analysis suggested that self selected gait
speed accounted for 47% of the variance of falls and stumbles
per 5000 right steps (p < 0.001).

**DISCUSSION**

Although younger, volunteers had considerable numbers of
falls and stumbles, emphasising the importance of studying a
control population. However, DM1 patients were far more
likely (approximately 10-fold) to fall and stumble, even
though they were active for only about 20% of the 24 hour
day compared with 28% in volunteers, walked more slowly,
and had substantially lower intensities of activity (table 1).

Some patients maintained high activity levels despite a
propensity to stumble or fall.

Volunteers stumbled or fell for obvious extrinsic causes.

Why patients fell or stumbled was often unclear; questioning
yielded no evidence for an association with preceding loss of
consciousness, and confidence did not appear relevant. As
expected, visual acuity was reduced and depression more
common in the patient group; both factors have been
associated with falling in the elderly. Patients who had falls were
less mobile by RMI, had slower self selected gait speeds
and higher depression scores, but did not have notably
different vision compared with those who did not.

Medication was not an obvious factor.

Distal weakness combined with knee and hip weakness
may particularly predispose to loss of pillar support following
minor sudden perturbations in stance. While adaptive
strategies may mitigate this risk these might fail if there is
mood or cognitive impairment: frontal lobe function and
visuospatial domains would perhaps be worthy of further
study. Only one patient was wearing ankle-foot orthoses in
this patient sample and this requires further systematic
investigation.

This study provides insufficient data to measure the
absolute likelihood of serious falls or resulting injury in
DM1 patients, but there seems no reason to believe that
either are less likely than in older people; comparison with
other neuromuscular disorders would be of interest. The
impact of osteoporosis is unclear in patients with muscle
disease, although the benefits of muscle activity on bone
mineral density are recognised. However, DM1 patients are
at relatively high risk of falls and stumbles and therefore of
injury. These events should form part of routine follow up
inquiry, and patients may benefit from a multifactorial
evaluation and multidisciplinary interventions similar to
elderly people.

**ACKNOWLEDGEMENTS**

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participated in the study. We thank Khaled Fallatah, Dr Iris Musa
(School of Healthcare Studies), and Ms Sarah Williams for assistance
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Dystrophy Support Group. MEB is supported by a grant from the
Wales Office for Research & Development. JF-M is supported by a
grant from the Muscular Dystrophy Campaign.

**Table 2** Stumbles and falls in the study populations

<table>
<thead>
<tr>
<th>Events 13 weeks</th>
<th>DM1 (n = 13; 7f)</th>
<th>Volunteers (n = 12; 7f)</th>
<th>p Value (Fisher’s exact test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>N who stumbled</td>
<td>10</td>
<td>9</td>
<td>1.00</td>
</tr>
<tr>
<td>No of stumbles</td>
<td>127</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>No who fell</td>
<td>6</td>
<td>2</td>
<td>0.202</td>
</tr>
<tr>
<td>No of falls</td>
<td>34</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>No who stumbled or fell 11</td>
<td>2</td>
<td>10</td>
<td>2</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Mean</th>
<th>SD</th>
<th>Mean</th>
<th>SD</th>
<th>p Value (Mann-Whitney test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Falls/person</td>
<td>2.62</td>
<td>4.56</td>
<td>0.25</td>
<td>0.62</td>
</tr>
<tr>
<td>Falls/5000 steps</td>
<td>0.076</td>
<td>0.167</td>
<td>0.002</td>
<td>0.006</td>
</tr>
<tr>
<td>Stumbles/person</td>
<td>9.62</td>
<td>19.25</td>
<td>2.17</td>
<td>2.37</td>
</tr>
<tr>
<td>Stumbles/5000 steps</td>
<td>0.130</td>
<td>0.219</td>
<td>0.019</td>
<td>0.021</td>
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<tr>
<td>Falls+stumbles</td>
<td>12.23</td>
<td>22.82</td>
<td>2.42</td>
<td>2.31</td>
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<tr>
<td>Falls+stumbles/5000 steps</td>
<td>0.206</td>
<td>0.288</td>
<td>0.021</td>
<td>0.021</td>
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</table>

<table>
<thead>
<tr>
<th>Classification of cause</th>
<th>No of subjects</th>
<th>p Value (χ²)</th>
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<tr>
<td>Extrinsic</td>
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<tr>
<td>Intrinsic</td>
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<tr>
<td>Non-bipedal</td>
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<td>0</td>
</tr>
<tr>
<td>Combination</td>
<td>0</td>
<td>3</td>
</tr>
</tbody>
</table>

*Excluding unclassified falls.

DM1, myotonic dystrophy type 1; f, female.
REFERENCES


