SHORT REPORT

Myotonic dystrophy: practical issues relating to assessment of strength

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Background: Myotonic dystrophy type 1 is a slowly progressive multisystem disease in which skeletal muscle involvement is prominent. As novel physical and pharmacological treatments become available, it is crucial to be able to measure their efficacy accurately.

Methods: 158 consecutive patients with myotonic dystrophy were assessed annually in a specialist muscle clinic. Strength was measured using both the Medical Research Council (MRC) scale and a hand-held dynamometer. Dynamometer readings were obtained from 108 normal subjects (controls).

Results: The movements showing the greatest rate of change in strength were ankle dorsiflexion and pinch grip. Both of these showed a decline of only 0.06 points/year on the MRC scale. Using a hand-held dynamometer, a change in strength of 1.18 kgN/year for women and 1.61 kgN/year for men was detected.

Conclusions: The MRC scale is unsuitable for detecting the small changes in strength seen in a slowly progressive disease such as myotonic dystrophy. Dynamometry provides a simple alternative that can give meaningful data over the duration of a typical clinical trial.

Myotonic dystrophy type 1 (DM1) is the most common inherited myopathy in adults, with a prevalence of about 8 in 100 000. It is a slowly progressive multisystem disorder, with skeletal muscle involvement being a prominent feature.^{1 2}

The role of exercise in ameliorating the relentless decline is uncertain, but there is some evidence suggesting that structured exercise programmes can slow or possibly reverse the decline in limb strength.^{3 4} The long-term goal is a treatment that corrects the underlying molecular disorder. Even then, such a treatment may only prevent further deterioration rather than reverse the existing deficits. The rate of change in muscle strength either during the natural course of the disease or after exercise programmes is very small and may be difficult to detect using traditional scoring methods.⁵

We compared the rate of change in strength of proximal and distal muscles as measured by the Medical Research Council (MRC) scale with that obtained using hand-held dynamometry to measure grip strength, in order to estimate the minimum time over which a change is detectable. Our observations have implications as to which clinical parameters should be measured during treatment trials in slowly progressive neuromuscular disorders such as DM1.

PATIENTS AND METHODS

Data on 158 consecutive adult patients (79 women, mean age 47 years, range 19–75 years; 79 men, mean age 43.6 years, range 12–75 years) with DM1 attending the routine annual

review at a specialist muscle clinic were analysed. The length of follow-up ranged from 1 to 8 years (mean 3.5 years). Patients were assessed by the same observer (DH-J) on each occasion, and the strength of the following movements graded according to the MRC strength scale: neck flexion and extension, shoulder abduction, wrist flexion and extension, pinch grip (between thumb and first digit), hip flexion and ankle dorsiflexion. At each evaluation, the results from previous assessments were not available to the assessor.

Patients were then asked to produce a maximum force on a handgrip dynamometer (Takei and Company, Ishioka City, Japan; range 0–100 kgN, scale graduation 0.5 kgN). Measurements were made with the patient standing, or sitting if unable to stand, with the dynamometer held at the side with arm straight, and the best of three attempts from each hand was recorded. Normal data were produced by asking 108 healthy volunteers (controls: 48 women, mean age 43.8 years, range 18–80 years; 60 men, mean age 44.5 years, range 18–80 years) to perform the same protocol.

RESULTS

MRC strength scale

Proximal muscle groups—for example, shoulder abduction—showed a mean decline in strength of only 0.01 and 0.02 MRC scale points/year for male and female patients, respectively. Even the most rapidly declining movement—namely pinch grip—showed a decline of only 0.05 and 0.06 points/year for male and female patients, respectively.

Dynamometry

Table 1 shows the results for patients and controls. To estimate the rate of change in grip strength expected in controls, linear regression analysis was carried out on grip strength versus age of the participant. This produced an estimated decrease of 0.21 kgN/year for women and 0.28 kgN/year for men.

We found that the mean grip strength at presentation (first clinic assessment) was 16.1 and 11.1 kgN for male and female patients, respectively, which differed significantly

Table 1 Dynamometry results of patients and controls

	Women		Men	
	DM1	Control	DM1	Control
Mean (kgN)	11.1	24.0	16.1	43.3
SE	0.88	0.96	1.5	1.1
Number	79	48	79	60
Change per year	1.18	0.21	1.61	0.28

DM1, myotonic dystrophy type 1.

Abbreviations: DM1, myotonic dystrophy type 1; MRC, Medical Research Council

from controls (43.3 and 24 kgN; $p = 0.05$). The mean rate of change in handgrip was a reduction of 1.18 kgN/year for female and 1.61 kgN/year for male patients.

DISCUSSION

As new genetic, pharmacological and physical treatments are developed for slowly progressive diseases such as DM1, there will be a need for accurate methods to assess their efficacy. Most clinical trials are conducted over weeks to months rather than over several years. Even if a new treatment were to completely halt progression of the disease, the difference in strength between treated patients and controls, even after a few years, would be small. Our data suggest that even if a trial were continued for a year and the treatment completely halted decline, using the MRC scale to assess the most clinically affected muscle groups (pinch grip and ankle dorsiflexion) would yield a change of only 0.06 points—which is clearly immeasurable. Other movements such as shoulder abduction and hip flexion show even smaller changes. This difficulty partly reflects the unsuitability of the MRC assessment scale, but more so the particular pattern of muscle weakness characteristic of DM1.

With manual muscle testing, a major problem is the range of strength covered by point 4 on the MRC scale. Subdivision to 4+ and 4– expands the scale but leads to problems with both intraobserver and interobserver variability. In this study, point 4 was not subdivided and the same clinician carried out all clinical assessments. Quantitative muscle testing using a hand-held myometer may seem to offer an alternative, but there are practical problems in assessing strong proximal muscles using the break technique.

DM1 presents a unique pattern of muscle weakness, well recognised by clinicians. Early on in the disease, there is weakness of the facial muscles, neck flexion, finger flexion and ankle dorsiflexion. This is often evident even in asymptomatic patients picked up during family screening (the exception being older patients with very small cytosine–thymine–guanine expansions). Conversely, proximal limb muscle strength (eg, shoulder abduction and hip flexion) remains normal even in relatively advanced disease. This is in striking contrast with the predominantly proximal weakness that characterises most of the acquired and inherited

myopathies. Manual muscle testing and quantitative muscle testing scores, which include many proximal muscles, are therefore unlikely to be sensitive to change in DM1.

Finger flexion, measured as grip strength, is a particularly attractive feature to measure in DM1. It is a movement affected early on in the course of the disease in virtually all patients, is a major cause of disability, and strength measurements can be made easily and reliably using simple and cheap equipment. There is little dependence on the examiner, and the results are highly reproducible. By using a hand-held dynamometer we found a reduction in strength that would be detectable after 6 months, a reasonable duration for a clinical trial. Our data imply that assessment of strength in future therapeutic trials should concentrate on specific areas, most notably grip strength.

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