ABSTRACT: We have assessed the reliability of hand-held myometry in 33 patients with spinal muscular atrophy (SMA), testing elbow flexion, handgrip, three-point pinch, knee flexion, knee extension, and foot dorsiflexion, and determining intraclass correlation coefficients (ICC). Interrater reliability was high for upper limbs, with an ICC of 0.92 for three-point pinch and 0.98 for elbow flexion and grip. For lower limbs interrater reliability was good with ICC >0.85 for all measures except foot dorsiflexion. Test–retest results were excellent with ICC >0.91 in all instances. Hand-held myometry is easily performed in SMA patients of various ages and muscle strengths, is a reliable measure of limb muscle strength, and can be used in longitudinal studies and clinical trials.

RELIABILITY OF HAND-HELD DYNAMOMETRY IN SPINAL MUSCULAR ATROPHY

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Spinal muscular atrophy (SMA) is the most common form of motor neuron disease in children and young adults with an incidence up to 10 per 100,000 live births.20,22 The disease is characterized by degeneration of anterior horn cells leading to progressive paralysis with muscle atrophy. Depending on the clinical type (Werdnig–Hoffmann, intermediate form, or Kugelberg–Welander syndrome, representing type I, II, or III, respectively),21 SMA causes early death or increasing disability in childhood or adulthood. Hence, measurement of muscle strength and residual motor function is important for assessing the effectiveness of potential treatments. The method of evaluation must be reliable, applicable to patients with a wide range of muscle strengths, easy to use in various clinical settings for multicenter trials, and ideally also suitable for use in clinical practice. The most common method of evaluating the strength of separate muscle groups is to use the Medical Research Council16 (MRC) scale that runs from 0 (no contraction) to 5 (normal power), in which ability to move against gravity is an important element. It is a convenient and inexpensive method, but depends on subjective evaluation, produces ordinal data, and requires extensive training to be able to correctly evaluate muscle strength, particularly when contractures limit the range of motion.8 Furthermore the MRC scale is not sufficiently sensitive to assess strength in very weak muscles, when movement is possible only if gravity is eliminated; sensitivity is also lacking for evaluating muscles that are powerful enough to overcome gravity but are still weak.2,29,30 Finally, the scale is characterized by a high level of intra- and interrater variability that compromises the results of clinical studies.8

To overcome these problems, quantitative muscle testing (QMT) is increasingly being used.2,8 Using a manual or fixed dynamometer,13 the strength of most clinically important muscle groups of the extremities can be measured and values in patients compared with those in the normal population. For following patients with neuromuscular disorders, strength measurement with a dynamometer is more reliable than grading using the MRC scale.2,10,29 A manual dynamometer, in particular, is simple to use even in wheelchair-bound patients, can detect small changes in muscle strength, and gives quantitative data over a continuous range. In addition, hand-held dynamometry compares favorably with fixed dynamometry in people with neuropathic

Abbreviations: CI, confidence interval; ICC, interclass correlation coefficient; MRC, Medical Research Council; QMT, quantitative muscle testing; SMA, spinal muscular atrophy

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Reliability of Dynamometry in SMA

MUSCLE & NERVE July 2002 65

weakness. It therefore appears as the most appropriate instrument for monitoring muscle strength in multicenter clinical trials.

Responsiveness and reliability of quantitative muscle strength testing have been demonstrated in amyotrophic lateral sclerosis where it is frequently used a primary outcome. QMT was used in studies of oxandrolone, and myoblast transfer as a primary efficacy measure in Duchenne muscular dystrophy. QMT has been employed as an outcome measure in clinical trials of therapeutic agents for spinal muscular atrophy, but reliability data were not provided. In addition, few studies on the reliability of QMT in SMA patients have been published, and their results are conflicting.

We designed a standardized protocol involving the use of a hand-held dynamometer to measure muscle strength in the SMA patients enrolled in a multicenter clinical treatment trial. Before the trial, we assessed inter- and intrarater reliability in the evaluation of muscle strength in patients (age range 5–65 years) with type II and type III SMA.

MATERIALS AND METHODS

Subjects. This study recruited all 33 patients with type II or type III SMA attending the Neuromuscular Unit of the Istituto Ortopedico Rizzoli, Bologna, Italy, from December 2000 to March 2001. The SMA diagnosis was confirmed genetically, and all patients gave their informed consent to participate. The study protocol was approved by the Institute’s Ethical Review Committee.

Testing. Maximum voluntary isometric contraction was assessed with a hand-held dynamometer (Type CT 3001, Citec, C.I.T. Technics BV, Groningen, The Netherlands). Six muscle groups were tested: handgrip; three-point pinch; elbow flexors; knee extensors; knee flexors; and foot dorsiflexors.

Patients were tested independently by two raters, a physiotherapist (rater A) and a neurologist (rater B), and subsequently retested by the physiotherapist. Each patient was tested in a single session with at least 20 min between each testing procedure, during which time the patient rested. To reduce the effects of patient bias, which may occur as a result of practice or fatigue, the order of testing (by the neurologist or the physiotherapist) was randomized. Test results were reported on forms given to the study coordinator and each rater was blinded to the results of the other’s testing. Test sessions were always held at about the same time of day in the same test room. Each muscle group was assessed on the patient’s preferred side only. The patient was instructed to increase to maximum exertion over 2–3 s without jerking, and to maintain this effort until the examiner gave the command to relax (3–5 s). The patient performed each movement three times with a 30-s pause between each. The highest score obtained was used for further analysis. The examiner held the myometer stable (“make” test) and the subject was asked to push as hard as possible against the myometer applicator, but with a slow build-up of force. If a patient complained of discomfort, additional padding was available to place on the applicator.

The elbow flexors were examined in the standard position; that is, supine, shoulder adducted, elbow flexed at 90°, forearm pronated, with the myometer just proximal to the wrist flexor crease. The testing positions for the other muscle groups were modified in order to test patients with reduced muscle strength, in those complaining of discomfort in the standard positions, and in those with contractures. To test pinch and full-fist grip, the subject was seated upright with the shoulder adducted, elbow flexed at 90°, forearm pronated, and wrist extended. No fixation was necessary. The examiner was seated or stood in front of the subject. For the pinch test the distal phalanx of the thumb was positioned under the applicator and the distal phalanges of the other two digits above the collar of the dynamometer, and the patient instructed to squeeze. For full-fist grip, subjects were seated with the dynamometer held perpendicularly in front of them by the tester and then required to squeeze the handgrip. To test lower limb muscles, the subject was seated on an examination couch or in a wheelchair with the hip and knee flexed at 90°, foot dorsiflexed at 90°, and told to grasp the edge of the couch or the arm rests with both hands. The Examiner sat in front of the subject and the dynamometer was placed on the anterior surface of the distal tibia just proximal to the ankle joint for testing the knee extensors, on the distal Achilles’ tendon for the knee flexors, and just proximal to the metatarsophalangeal joints (dorsal surface) for the foot dorsiflexors.

Statistical Analyses. Intraclass correlation coefficients. To assess inter- and intrarater reliability, we calculated intraclass correlation coefficients (ICCs). ICC scores were interpreted conventionally: <0, poor agreement; 0–0.20, slight agreement; 0.21–0.40, fair agreement; 0.41–0.60, moderate agreement; 0.61–0.80, substantial agreement, and 0.81–1, almost perfect agreement. The 95% confidence intervals were determined using the bootstrap method.

Bland–Altman plots. The differences between the measurements made by the two raters were plot-
tested against their means (Bland–Altman plots) to check whether the measurement error was independent of score magnitude. Data were analyzed using the SAS-PC software package (SAS Institute, Inc., Cary, North Carolina).

RESULTS

There were 17 men and 16 women, aged 5–65 years (mean 22.9 ± 11.4 years). Among them, 21 (63.6%) had type II, and 12 (36.4%) had type III SMA. At the time of examination, 5 of the 12 patients with type III SMA were still able to walk. None of the patients experienced joint or soft tissue discomfort during testing. The means, standard deviations, and ranges of measured muscle strength for the three evaluations are shown in Table 1. No consistent pattern of decrease or increase between the three evaluations was observed. Analysis of the mean values of the first, second, and third measurements of tester A1 for all patients showed that the first measurement was the highest on two occasions (grip and knee extension), the second measurement was the highest on three occasions (elbow flexion, knee flexion), and the third measurement was highest on one occasion (foot dorsiflexion). A steady decrease from the first to the third measurement was found only twice (grip and knee extension).

**Intraclass Correlation Coefficients.** Test–retest analysis (Table 2) showed high intrarater reliability, with ICC scores ranging from 0.94 to 0.98 on the upper limbs, and from 0.91 to 0.97 on the lower limbs. Interrater agreement was also excellent for upper limb testing, with ICC scores ranging between 0.92 and 0.98, and good for the knee tests, with ICC scores between 0.88 and 0.95, but less satisfactory for foot dorsiflexion (0.69). Six of the 33 patients were below the age of 12 years, but analysis of variance showed no effect of age (below 12 versus above 12 years) on muscle strength (data not shown).

**Bland–Altman Plots.** Bland–Altman plots, in which the difference between the two measurements are plotted against their mean value, are shown for upper limb and lower limb muscle groups in Figures 1 and 2, respectively. Visual inspection shows that better results were obtained for upper limb muscle groups and for intrarater testing, whereas lower limb points (Fig. 2) deviated more from the mean than those for the upper limb (Fig. 1). For foot dorsiflexion, there were marked differences in test–retest score in three instances: in one patient the first result was 20 N (Newtons) and the retest 49 N; in another the first result was 46 N and the retest 83 N; and in the third the first result was 68 N and the retest 102 N. The latter result is shown in Figure 2 (bottom left), as the point at 85 (mean of 68 and 102) and −34 (difference between 68 and 102). In three instances, also, dorsiflexion measurements differed considerably between the two raters and hence the points representing them (Fig. 2, bottom right) were distant from the line of the mean. In one patient, rater A measured 5 N and rater B 39 N; in another patient, rater A measured 20 N and rater B 114 N; and, in a third patient, rater A measured 46 N

### Table 1. Muscle strength in 33 spinal muscular atrophy (SMA) patients.

<table>
<thead>
<tr>
<th>Muscle group</th>
<th>Tester A (1)</th>
<th>Tester A (2)</th>
<th>Tester B</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hand grip</td>
<td>12.1 ± 16.4</td>
<td>11.1 ± 14.4</td>
<td>11.5 ± 16.3</td>
</tr>
<tr>
<td>Elbow flexion</td>
<td>23.2 ± 27.3</td>
<td>22.2 ± 25.0</td>
<td>23.7 ± 29.3</td>
</tr>
<tr>
<td>Three-point pinch</td>
<td>12.6 ± 13.7</td>
<td>14.1 ± 15.8</td>
<td>12.2 ± 13.5</td>
</tr>
<tr>
<td>Knee extension</td>
<td>4.9 ± 5.9</td>
<td>4.8 ± 6.4</td>
<td>3.3 ± 7.2</td>
</tr>
<tr>
<td>Knee flexion</td>
<td>15.9 ± 14.5</td>
<td>16.2 ± 13.7</td>
<td>15.5 ± 16.1</td>
</tr>
<tr>
<td>Foot dorsiflexion</td>
<td>17.0 ± 22.8</td>
<td>19.6 ± 27.4</td>
<td>23.2 ± 34.9</td>
</tr>
</tbody>
</table>

Data are mean ± standard deviation (range) of the maximal voluntary isometric contraction strength expressed (in Newtons) in 33 SMA patients as measured by tester A (twice) and by tester B (once) using a hand-held dynamometer.

### Table 2. Reliability of strength assessment using a hand-held dynamometer in 33 SMA patients.

<table>
<thead>
<tr>
<th>Muscle group</th>
<th>Intrarater ICC (95% CI)</th>
<th>Mean (95% CI) score difference</th>
<th>Interrater ICC (95% CI)</th>
<th>Mean (95% CI) score difference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hand grip</td>
<td>0.97 (0.93–0.98)</td>
<td>0.93 (−0.4–2.2)</td>
<td>0.98 (0.95–0.98)</td>
<td>0.54 (−0.6–1.7)</td>
</tr>
<tr>
<td>Elbow flexion</td>
<td>0.98 (0.95–0.99)</td>
<td>1.00 (−0.9–2.9)</td>
<td>0.98 (0.96–0.99)</td>
<td>−0.45 (−2.3–1.4)</td>
</tr>
<tr>
<td>Three-point pinch</td>
<td>0.94 (0.89–0.97)</td>
<td>−1.48 (−3.1–0.19)</td>
<td>0.92 (0.84–0.95)</td>
<td>0.48 (−1.4–2.4)</td>
</tr>
<tr>
<td>Knee flexion</td>
<td>0.97 (0.93–0.98)</td>
<td>−0.21 (−1.4–1.0)</td>
<td>0.95 (0.90–0.97)</td>
<td>0.48 (−1.2–2.2)</td>
</tr>
<tr>
<td>Knee extension</td>
<td>0.93 (0.85–0.96)</td>
<td>0.12 (−0.7–0.9)</td>
<td>0.88 (0.77–0.94)</td>
<td>1.63 (0.5–2.7)</td>
</tr>
<tr>
<td>Foot dorsiflexion</td>
<td>0.91 (0.83–0.95)</td>
<td>−2.60 (−6.2–1.0)</td>
<td>0.69 (0.46–0.83)</td>
<td>−6.24 (−14.3–1.8)</td>
</tr>
</tbody>
</table>

CI, confidence interval; ICC, intraclass correlation coefficient; SMA, spinal muscular atrophy.
FIGURE 1. Intra- and interrater agreement for upper limb muscle strength testing expressed by Bland–Altman plots. The solid line is the mean of the differences in all subjects; the dotted lines are 2 SD above and below the mean. On the left are intrarater plots, and on the right interrater plots.
FIGURE 2. Intra- and interrater agreement for lower limb muscle strength testing expressed by Bland–Altman plots. The solid line is the mean of the differences in all subjects; the dotted lines are 2 SD above and below the mean. On the left are intrarater plots, and on the right interrater plots.
and rater B 131 N. For one patient the knee extension measurement differed considerably between the two raters (Fig. 2, middle left) with rater A measuring 12 N and rater B only 1 N.

Four of these seven largest differences concerned measurements in two patients, neither of whom were children (a 64-year-old woman and a 32-year-old man). We found no tendency for one rater to systematically over- or underestimate maximum muscle strength compared to the other.

The Bland–Altman plots indicate that the size of the difference between the two raters was independent of muscle strength for all muscle groups except the three-point pinch. For this muscle group, interrater differences were greater for strength scores over 15 (Fig. 1, right column, bottom), suggesting that these measurements are less reliable in individuals with greater strength.

**DISCUSSION**

This study has shown that maximum voluntary isometric contraction measured by a hand-held myometer provides reliable results in patients, of various ages and strengths, with SMA. Intrarater results were excellent with ICC >0.90 in all cases; interrater reliability was excellent for upper limb strength, and good for lower limb measurements, with ICC >0.85 for all tests except foot dorsiflexion. As expected, intrarater reliability was higher than interrater reliability for all muscle groups. Previous studies have shown that quantitative muscle testing is reliable with various types of equipment, in normal subjects, and patients with neuromuscular disorders, particularly those with Duchenne muscular dystrophy, reports on patients with SMA have been preliminary or conflicting.

In this study, as in others, interrater reliability was less satisfactory for ankle dorsiflexion (ICC = 0.69). Escolar et al. obtained reliable QMT in 12 children with Duchenne muscular dystrophy for biceps and grip strength (ICC >0.9), and for quadriceps strength (>0.8), but not for foot dorsiflexion (ICC <0.75). Phillips et al. also obtained good results in adult patients, with coefficients >0.85 in 17 muscle groups for both intrasession and intersession reliability, with the exception of ankle dorsiflexor results.

Several factors have been suggested as contributing to the poor reliability of foot dorsiflexion testing. The main factor is the short lever arm used for testing foot dorsiflexion and the difficulty of aligning the myometer transducer precisely in the line of force exerted by the subject. Foot dorsiflexion is particularly problematic in patients with neuromuscular disorders because of the presence of ankle contractures, which often render myometer positioning difficult and inconsistent. Moreover, as foot dorsiflexion is measured in the sitting position, it is not possible to eliminate gravity. Given the poor reliability of foot dorsiflexion, we suggest not using it as a response measure in clinical trials involving patients with neuromuscular disorders.

We found that the error of measurement was independent of the magnitude of the score for most muscle groups. The only exception was the three-point pinch, where the results were less consistent for scores greater than 15. The pinch test requires more careful positioning of the dynamometer than the grip test, which was associated with high inter- and intrarater ICCs in our study. Both these tests evaluate flexor muscles innervated by the same lower cervical segments, and measurement of the pinch does not provide information additional to that provided by the grip test and could therefore be eliminated.

In our study population the weakest muscles were those concerned with knee extension. This contrasts with the situation in normal children and adults, where the knee extensors are the strongest muscles. This finding is consistent with the well-known segmental distribution of muscle weakness that characterizes SMA patients, particularly those with type III disease, in whom proximal muscles are weaker than distal muscles and knee extensors are weaker than knee flexors.

Little information is available of the use of QMT in patients with neuromuscular disease, and contrasting results have been reported by different groups. One study group, using a fixed myometric system, succeeded in detecting differences among patients with Duchenne muscular dystrophy that could not be detected by manual muscle testing, but not among the individual muscle groups, although reliability data were not presented. By contrast, the Tufts quantitative neuromuscular examination has been shown to have good reproducibility and to be able to identify large differences in mean strengths of seven muscle groups of children with Duchenne muscular dystrophy, supporting the idea that QMT provides objective measurements in children with varying force abilities.

Data have been collected for normal children and adults using hand-held dynamometry. This will allow comparison between patients with various neuromuscular diseases and appropriate gender- and age-matched normal data to accurately identify the presence of weakness.
in randomized double-blind clinical trials in patients with amyotrophic lateral sclerosis\textsuperscript{12} and Duchenne muscular dystrophy.\textsuperscript{9,17} However, rigorous training, and high standards are recommended to reduce the variability of results to an acceptable level.\textsuperscript{12} Since interrater reliability is lower than intrarater reliability, and does not seem to improve with practice,\textsuperscript{12} it has been recommended that individual patients should be assessed by the same rater throughout the study,\textsuperscript{10} particularly in multicenter trials.

In our experience, hand-held dynamometry provides reliable measures of limb muscle strength in SMA patients of different ages and strength and can be used in longitudinal studies and clinical trials of the disease.

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