

Is a reduced muscle test sufficient to give an overall impression of muscle function in spinal muscular atrophy II ?

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Background

Spinal muscular atrophy II (SMA II) is an inherited neuromuscular disease characterized by degeneration of the spinal cord motor neurons. The child with SMA II achieves the ability to sit independently but not to stand and walk independently. Overall muscle strength is limited, but upper limbs are usually stronger than lower limbs.

Regular assessments of muscle function are necessary to determine the progression of the disease and to plan rehabilitation. Manual muscle test (MMT) is useful for this purpose, but the test is time consuming and often cumbersome for severely disabled people.

Aim

The aim of this study was to examine to which degree a reduced MMT of the upper limbs is sufficient to give an overall impression of muscular function in a person with SMA II, and to study the change of muscle function over time measured by a full MMT (32 muscle groups) and a reduced MMT (14, 10, or 6 muscle groups).

Conclusion

The study indicates that a muscle test of the upper limbs is sufficient to give an overall impression of muscle strength in a person with SMA II. A percentage of muscle strength for the upper limbs or forearms could be a relevant measurement to illustrate development over time.



Methods

All persons ≥ 5 years of age registered in the National Rehabilitation Centre for Neuromuscular Diseases with a clinically and genetically confirmed diagnosis of SMA II ($n=67$) were invited to participate in the study.

The participants were assessed with MMT.

The MMT score was transformed to a ten point scale (CIDD protocol) and the percentage of total muscle strength (32 muscle groups), the percentages of muscle strength for the upper limbs (14 muscle groups), the forearm (10 muscle groups) and the hand (6 muscle groups) were calculated.

Correlations between muscle tests were calculated with Spearman Rank order correlation coefficient.

Descriptive statistics were used to illustrate distribution of data.

Muscle strength for persons ($n=20$) measured by MMT with intervals of six to twelve years were indexed to study change in muscle strength over time.

Results

Fifty-five persons participated in the study. Mean age was 23.6 years (5.1-69.5). Correlations between muscle tests were significant. Table 1. A reduced muscle test of the arms expressed a larger variation and discrimination among the participants than a total muscle test.

Fig. 1 and fig.2.

When muscle strength was measured over time a reduced muscle test showed a greater loss of muscle strength than a total muscle test. Fig. 3.

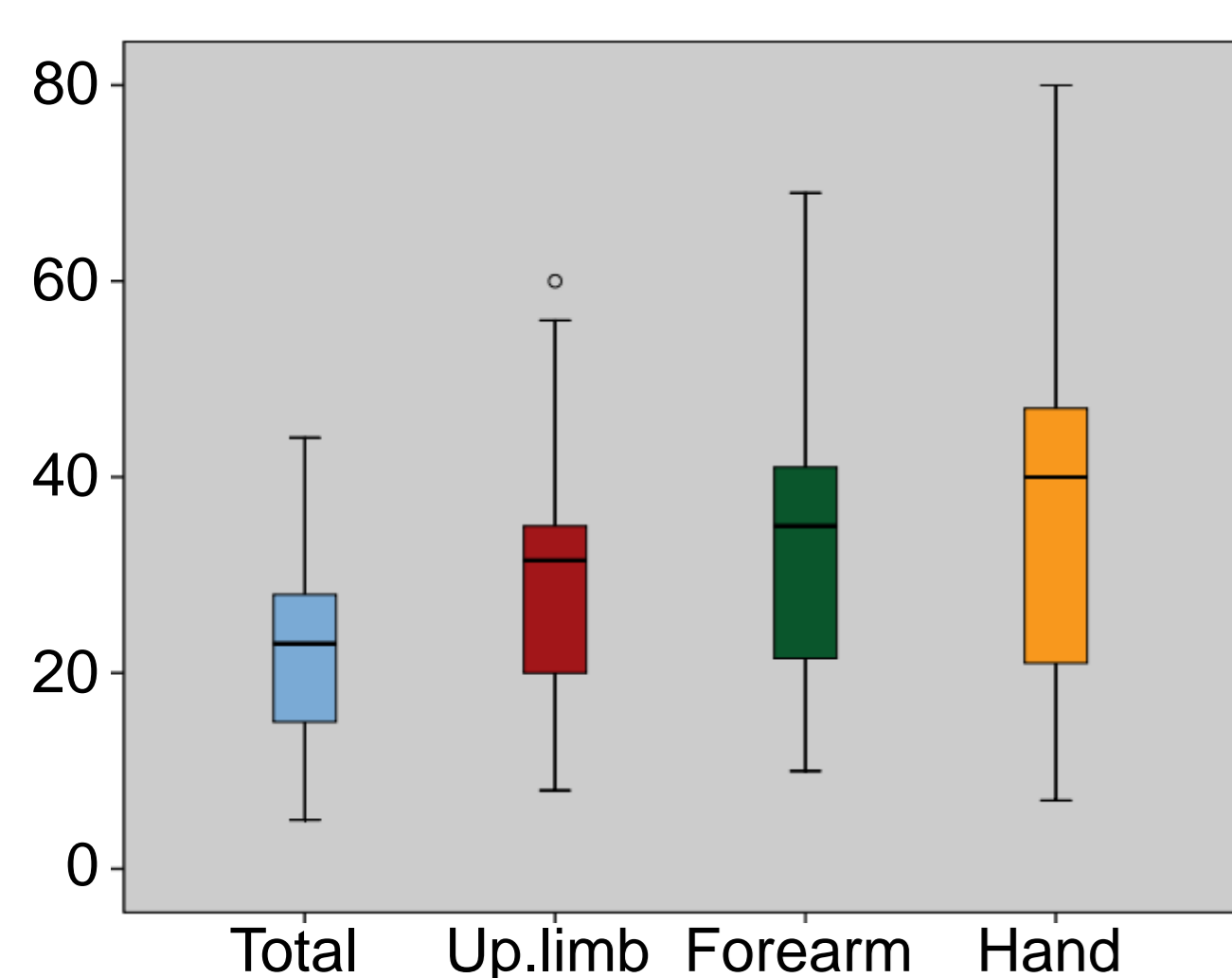
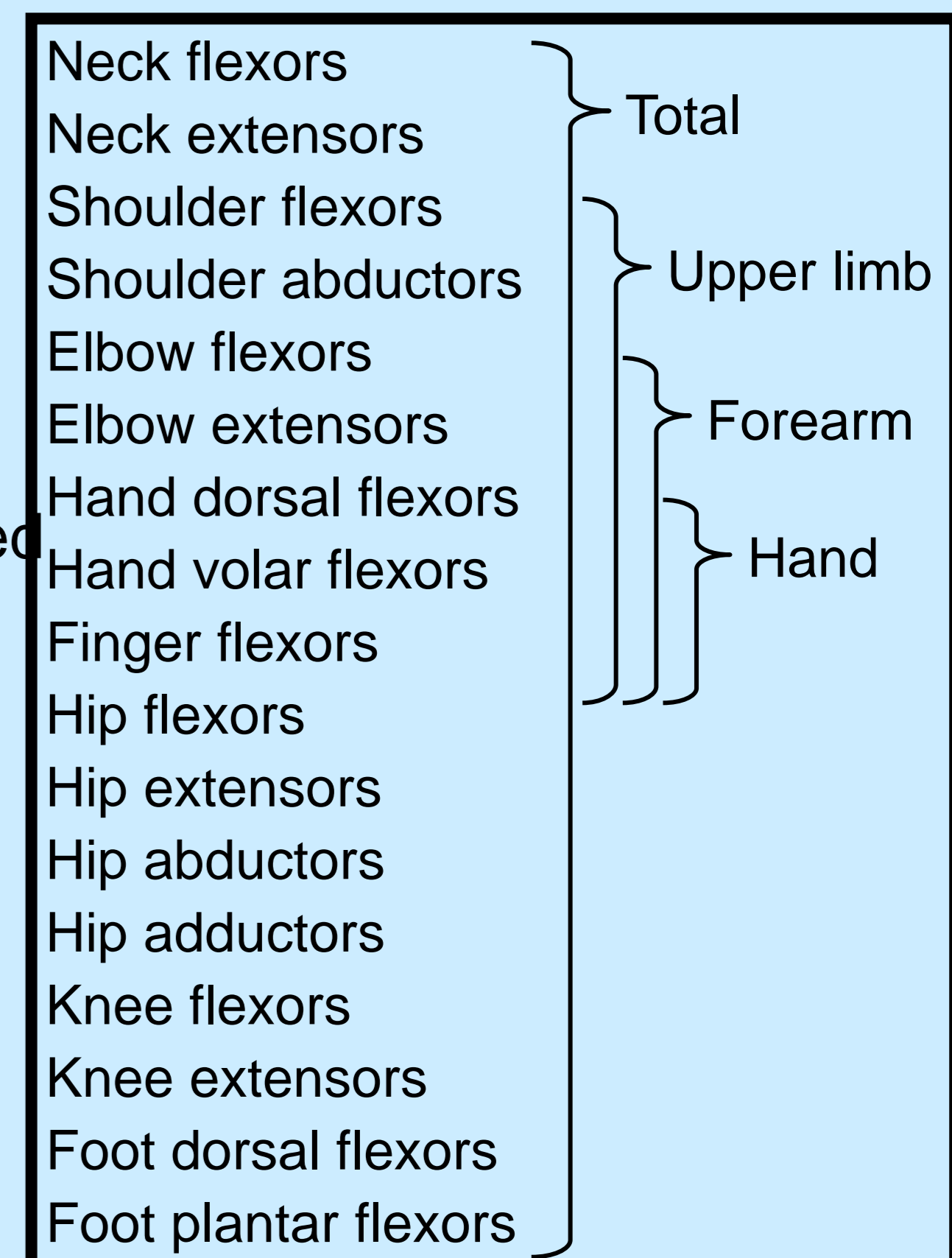


Fig. 1
Muscle strength distribution for 55 persons, when muscle strength was calculated as percentage of normal value. Total (32 muscles), Upper limb (14 muscles), Forearm (10 muscles), Hand (6 muscles)

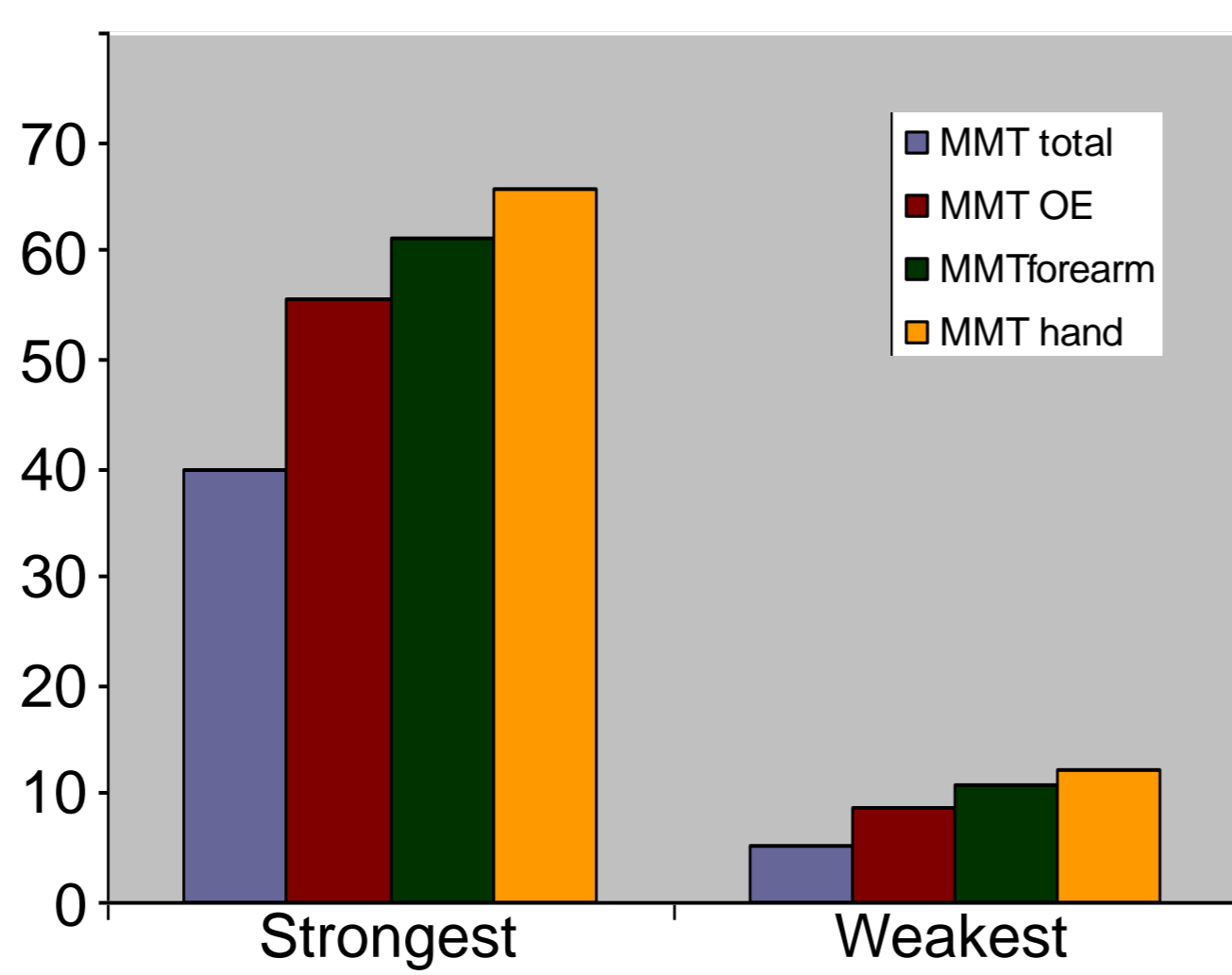


Fig. 2
Mean values for the three strongest persons and the three weakest persons, concerning total muscle strength and muscle strength in upper limb, forearm and hand

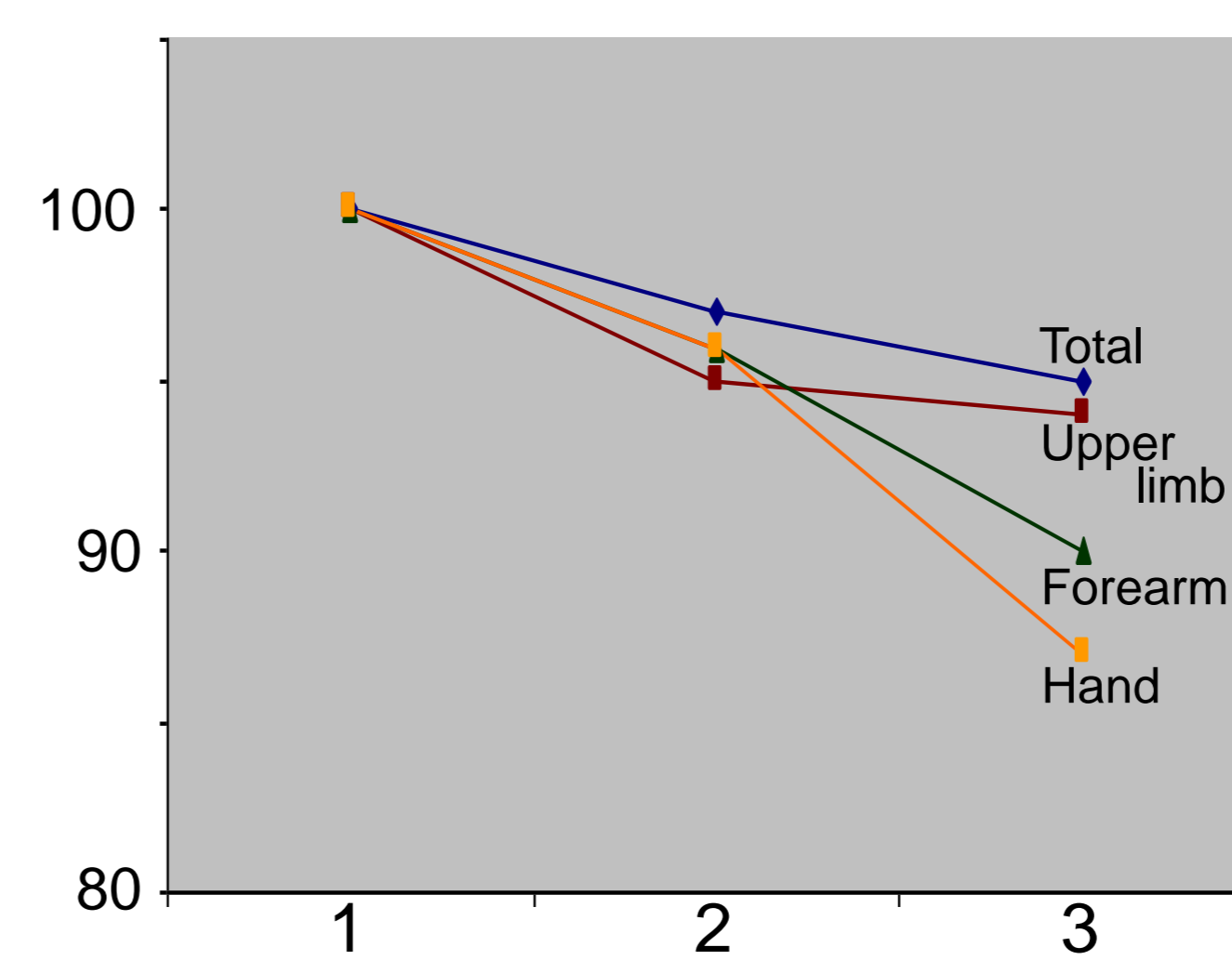


Fig. 3
Index for mean value of muscle tests over a period of twelve years. Time between tests is six years. Eighteen persons were examined with two or more muscle tests over a period of twelve years.

	Total	Upper limb	Forearm
Total (32 muscles)	1.000		
Upper limb (14 muscles)	.935	1.000	
Forearm (10 muscles)	.921	.993	1.000
Hand (6 muscles)	.875	.851	.971

Table 1
Spearman's rho correlation between percentage of normal value for 55 individuals. correlation is significant at the 0.01 level (2-tailed).