How well is functional ability related to muscle strength in spinal muscular atrophy II ?

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Background

Spinal muscular atrophy (SMA) is an inherited neuromuscular disease characterized by degeneration of the spinal cord motor neurons. According to the diagnostic criteria there are three types of SMA.
The clinical spectrum ranges from massive hypotonia and weakness to only mild weakness. The severity of weakness generally correlates to age of onset. SMA II is thus characterized with an onset from six months of age and the ability to sit unsupported.
Physical function in SMA II is assessed with a number of scales such as Hammersmith Functional Motor Scale (HFMS), EK scale, Brooke upper limb scale and manual muscle test (MMT) recorded as MRC and dynamometric measure.
Former studies have shown a close relationship between EK scale and Brooke upper limb scale but little is known about the relationship among the other assessment scales in this population.

Aim

The aim of this study was to investigate how functional ability assessed with HFMS, EK and Brooke related to muscle strength measured with MMT and dynamometer in the Danish population of people with SMA II ≥ 18 years.

Conclusion

Functional tests and muscle tests correlated significantly when used in a study of adult people with SMA2. The strongest relationship was found between EK and muscle tests, and Brooke and muscle tests.
Dynamometry measure with the CITEC dynamometer is complicated because persons with very little muscle strength cannot activate the dynamometer.
The activities on the HFMS, which is originally developed for children, were too difficult to complete for our population of adults with SMA2.

Methods

All persons ≥ eighteen years (n = 37) registered with RehabiliteringsCenter for Muskelsvind with a diagnosis of SMA II (clinically and genetically confirmed) were invited to participate in the study. Functional ability was assessed with HFMS, Brooke, EK. Muscle strength was assessed with MMT and a hand-held dynamometer (CITEC).
MRC score was transformed to a ten point scale and the overall percentage of muscle strength (40 muscles) and the percentage of muscle strength for the upper limbs (16 muscles) were calculated. Dynamometry (N) was measured bilaterally three times at elbow flexion, elbow extension and the grip. Sum of highest score was used to calculate Dynamometry total. Sum of highest score for right and left elbow flexion was used to calculate dynamometry elbow flexion.
Correlation was calculated with Spearman Rank order correlation coefficient at a significant level (two-tailed) of 0.01.

Results

Twenty nine persons participated in the study. Mean age was 34,7 years (18,1-69,5). Eighteen persons couldn't perform any task on the Hammersmith scale and scored zero. The mean percentage of overall muscle strength was 20 % (7-45).
The scores for functional ability (Brooke, HFMS, EK) and MRC are illustrated in fig. 1

Eight persons hadn't enough muscle strength to overcome any of the thresholds from the dynamometer. Elbow flexion could be measured in the remaining 21 persons whereas elbow extension could be measured in fourteen persons, and grip could be measured in eighteen persons.

There was a significant relationship between functional ability and muscle strength. Table 1.

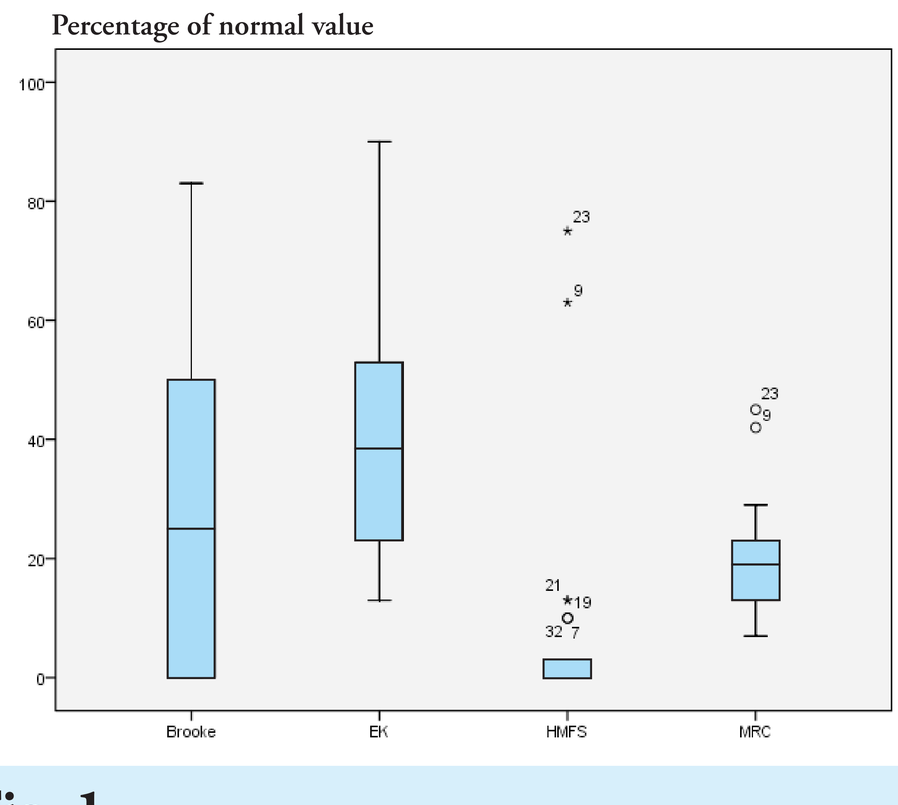

Fig. 1
Percentage of maximal ability and distribution of data for Brooke (30 persons), HMFS (30 persons), EK (30 persons) and MRC – 40 muscles (29 persons).
For each measurement scores was transformed to percentage of normal value.

	1 EK	2 HMFS	3 Brooke	4 MRC overall	5 MRC upper limb	6 Dyn total
1 EK						
2 HMFS	-0.62					
3 Brooke	0.93	-0.66				
4 MRC overall	-0.87	0.60	-0.91			
5 MRC upper limb	-0.86	0.56	-0.92	0.96		
6 Dynamometry total	-0.89	0.53	-0.93	0.92	0.93	
7 Dynamometry elbow flexion	-0.87	0.51	-0.90	0.89	0.89	0.99

Tabel 1
Correlation (Spearmanns rho) between measurements of functional ability and muscle tests.
Each correlation is significant at the 0.01 level

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