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Does muscle strength deteriorate in Spinal muscular atrophy type II and III?

15-year follow-up study in 23 patients with SMA II and 7 patients with SMA III

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Aim

The aim of this study is to evaluate muscle strength and physical functions over a period of at least 15 years in patients with confirmed SMA types II and III.

Conclusion

In this study we showed that physical function and muscle strength in the upper limbs as measured by MMT deteriorates in SMA II patients over time. A decrease in muscle strength of upper limbs was also shown in SMA III patients.

Introduction

Spinal muscular atrophy is associated with degeneration of the motor neurons in the spinal cord leading to muscle weakness. Although it is generally accepted that patients with Spinal muscular atrophy (SMA) types II and III loose motor abilities over time, it is still debated whether muscle strength deteriorates, and what outcome measures and methods should be used to assess this. Some studies have indicated that muscle strength as measured by manual muscle test (MMT) declined over time, but other studies could not demonstrate deterioration in muscle strength over time, when muscle strength was assessed by quantitative methods.

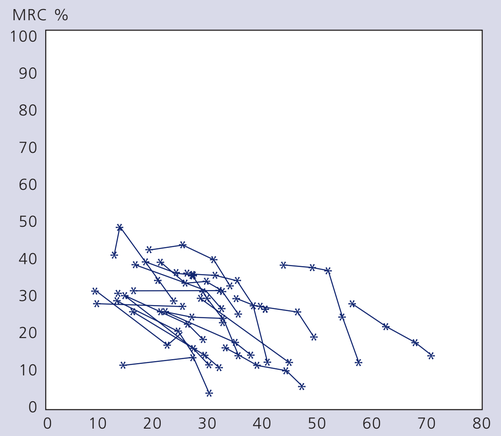


Fig. 1
Muscle strength over time in upper limbs – SMA II patients (n=21).
Change over time (p <0.0001)

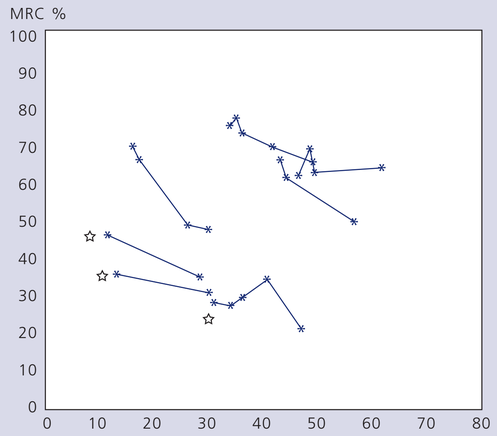


Fig. 2
Muscle strength over time in upper limbs – SMA III patients (n=7).
☆= non ambulant patients
Change over time (p = 0.0156)

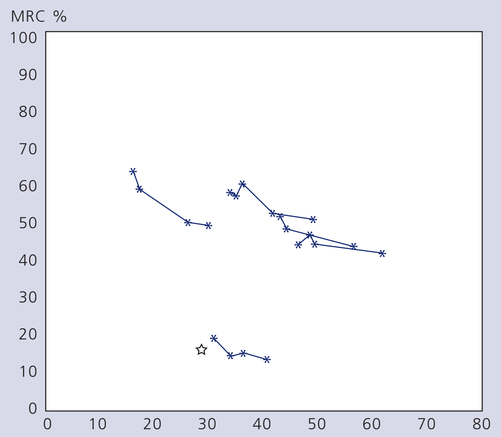


Fig. 3
Muscle strength over time in lower limbs – SMA III patients (n=5).
☆= non ambulant patient.
Change over time (p = 0.0625)

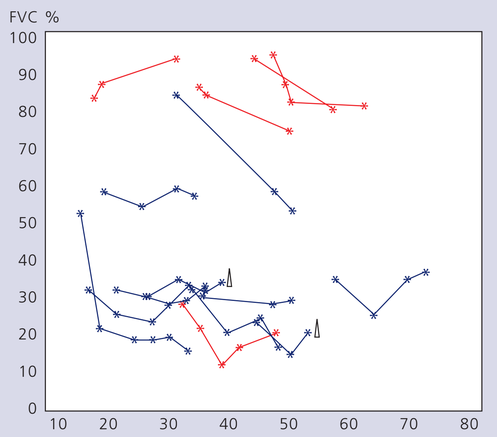
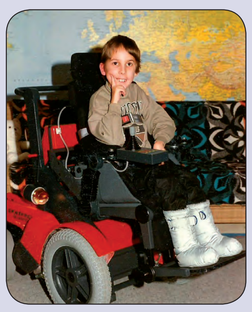


Fig. 4
FVC% over time in SMA II patients (n = 10/blue) and SMA III patients (n = 5/red).
⍋= patients with tracheostomy at last assessment.
Change over time (p = 0.2764, p = 0.1875)

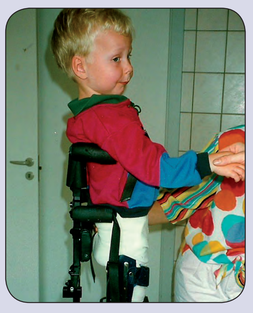
Materials and methods

Data from 23 patients with SMA II and 7 patients with SMA III - compiled prospectively at the National Rehabilitations Centre for Neuromuscular Diseases since the early nineties - were reviewed. Inclusion criteria were at least 15 years interval between first and last assessment.
Upper limb function was evaluated by means of Brooke upper limb scale and Manual Muscle Test (MMT). In ambulant SMA III patients, MMT of the lower limbs was also evaluated. MRC score was modified to a 0-10 score and MRC score % was calculated.
In non-ambulant patients, functional ability was evaluated by means of the EK scale.
Forced vital capacity was measured by means of spirometry. In patients with tracheostomy FVC from the assessment before the operation was recognized as the last assessment. When calculating FVC % predicted, only data from patients ≥ 14 years at first assessment were used.

Differences within groups were calculated by Wilcoxon signed rank test.


Patient 2

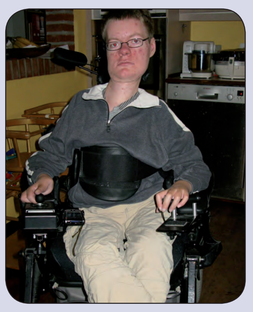

Patient 4


Patient 5


Patient 7


Patient 2


Patient 4


Patient 5


Patient 7

Results

Median age at entry was 15 years (7-53) in patients with SMA II and 31 years (11-47) in patients with SMA III.
Upper limb function as measured by Brooke scale declined in SMA II patients (p<0.0001), this was not the case for SMA III patients. Muscle strength in the upper limbs deteriorated over time in both SMA II patients (Fig. 1) and SMA III patients (Fig. 2), whereas a statistically decline in muscle strength in the lower limbs did not occur in this small group of SMA III patients (Fig. 3).
Physical function as measured by EK scale deteriorated in SMA II patients (p<0.0001); no decline was shown in the three non-ambulant SMA III patients.
FVC was evaluated in ten SMA II patients and five SMA III patients. Two SMA II patients had tracheostomy at the last assessment. There was no statistically decline in FVC % over time in neither SMA II patients nor SMA III patients (Fig. 4).

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