



Strengthening in Children with Spinal Muscular Atrophy (SMA) type III. Sense or Nonsense?



A. Oehrli-Thijs, R.-I. Hassink, J. Scheurer



Zentrum für Entwicklungsförderung und pädiatrische Neurorehabilitation
der Stiftung Wildermeth Z.E.N. , Kloosweg 22, CH-2502 Biel

Introduction

In children with **SMA type III**, the second motor neurons in the medulla spinalis are reduced. Therefore the impulses from the cortex are not forwarded to the muscles. We don't have conclusive answers about the success of **muscle strength training and aerobic exercise programs** for children with this disease.

We tried to answer the question, whether an improvement of muscular strength and physical fitness can be achieved by intensive training, at least 3 times/week, during 3 months, instead of continuous long term training, 1 time/week.

Methods

Case

Girl, 9 years old, SMA type III

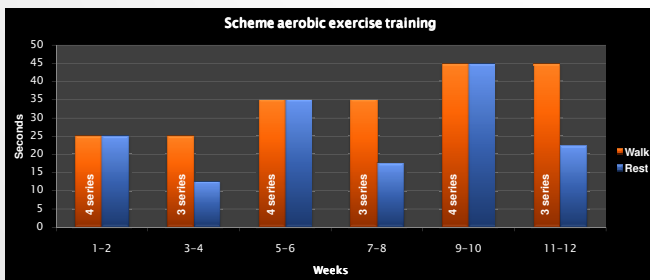
Training intensity, 3 times/week during 12 weeks

Considered recovery time: 72 hours

Capacity: self-manageable walking time: 34 seconds, distance 10 meter

Muscle strength of the relevant groups: M2

Aerobic exercise program



Due to the limited ability to walk longer than 34" it should be anaerobe training in the phosphate-lactate system, at 80-90% of the possible training intensity. However, we assumed, regarding the possibilities of the girl, it would rather be **aerobe training at 60-70%** of the possible training intensity

Muscle strength training

Methods with low level power use

- Moderate to high repetitions (4-12 rep, 3 series , 120")
- Low capacity of the patient during the training cycle
- The fatigue shouldn't influence the sensomotor achievement
- Concerning the moderate coordination we admitted the compensatory mechanism
- Methods of differentiated strength training
- Training method → therapeutic resistance
- Guidelines → general methodical principles of the AAP, the ACSM and the AOSSM
- No exercise machines

Results

In our case, the patient was able to increase her self-manageable walk distance at **240%**. Not included was the performance of the movement quality. We allowed her to move compensatory. Parents and patient were very satisfied with the result.

COPM satisfaction	6	→	8
COPM performance	5	→	6
COPM importance	8	→	8

Improvement of quality of life

The 1st follow-up three months after the intensive phase showed a reduction of 20% of the accomplished capacity.

Background

Muscle strength training in children means:

- No increase of muscle mass
- improving intramuscular coordination
- improving intermuscular coordination
- By training the interaction between agonist and antagonist is influenced in an optimal way
- Increase of effect by mounting the **stimulation time** (younger children) or the **stimulation level**

Aerobic exercise programs in children

- Effective realization by using special tests (Shuttle Run/ ½ Bruce etc.)
- Define the optimal heart rate for the training
- Good definition of the intensity, type of exercises, progression rate, frequency, duration of exercise session and of the entire program is very important

Discussion

- No subjective signs of exposure
- Can we assume, that by the specific strain of the patient, the energy level is different to normal?
- To assume: aerobe training = aerobe changes?
- Question:**
Would an increase of the training program (intensity, duration, frequency etc) support a positive effect?

By our means, it was not possible to investigate the reasons of the change.

More research is needed!

At what level the changes have taken place?

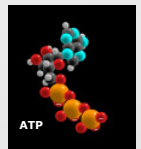
Energy supply?

- Higher lactate supply?
- Higher ATP supply?
- Better oxygen saturation?

Difference in muscle structure?

- Actin-myosine** cross-bridges?
- Regeneration of **muscle fibers**?
- Restructuring** of muscle fibers (type I/type II)?

Intermuscular coordination?



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Conclusion

This case tried to show that **muscle strength training and aerobic exercises** in children with **SMA type II** can bring a significant improvement, also in **quality of life**. However, it has yet to be shown, whether it makes a difference when patients train more frequently or with an increase of the program as described. Still, more research is to be done. And, the **individuality** of each patient influences the decisions to be made.