



Physical performance of children with Prader-Willi Syndrome following a 10-week sports therapeutic intervention

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Summary

Prader-Willi Syndrome (PWS) is a complex neurogenetic and the best characterized form of syndromic disorder, manifesting itself in markedly increased body fat as well as reduced muscle mass. Children with PWS show a reduced level of physical activity and impaired motor abilities. In a cross-over design the effects of a 10-week training intervention (TI) and a 10-week unstructured exercise program (control intervention (CI)) on motor abilities and body composition of 14 children with PWS were examined. Most parameters regarding local muscle strength increased statistically significant in children with PWS after TI. No significant changes of any parameter could be observed following CI. The results demonstrate that a specific TI has positive effects on physical performance of children with PWS and seems more effective than an unstructured training intervention. Further studies have to develop more detailed recommendations regarding the intensity and duration of exercise interventions for children with PWS in order to integrate targeted physical training into a comprehensive treatment management and therefore to enhance the children's quality of life.

Introduction

Prader-Willi Syndrome (PWS) is a complex multisystemic, neurogenetic disorder caused by a defect on the paternal chromosome 15. With a prevalence between 1:10.000 and 1:30.000 PWS affects both genders at similar rates. PWS manifests itself in, among others, endocrine disruption, behavioral and cognitive deficits, an abnormal body composition of markedly increased body fat as well as reduced muscle mass and is associated with highly developmental delays [1]. Several studies demonstrated an overall reduced level of physical activity and impaired motor abilities in children with PWS including reduced muscle strength, muscle tension and cardiovascular fitness as well as difficulties regarding balance and postural stability [2,3]. Physical exercise programs are an effective tool to prevent the severity of comorbidities of several diseases and initial studies demonstrated that children with PWS profit equally from physical training as healthy children [3]. Thus, aim of this study was to investigate the effects of a 10-week sports therapeutic intervention on physical performance of children with PWS.

Methods

Fourteen children with PWS (age: 16±2 years; body weight: 58±8 kg; BMI: 23.6±4.5 kg/m²; body fat: 23±4%) of two separated living groups were included in a cross-over design, whereby all 14 children completed TI as well as CI. By doing so, the effects of a 10-week training intervention (TI) and a ten-week control intervention (CI) on aerobic fitness (6-minute walking test), balance (one-leg stand), mobility (toe touch test) and muscle strength (sit-up, push-up, standing long jump) of children with PWS were examined. For the TI, children with PWS performed a specific training with its main focus on strengthening exercises (Fig. 1). For the CI, the common unstructured exercise program of the living groups, including walking and gymnastic sessions, was used. Both, TI and CI, were implemented as group intervention two times a week for a duration of 60 minutes and guided by caretakers of the living groups.

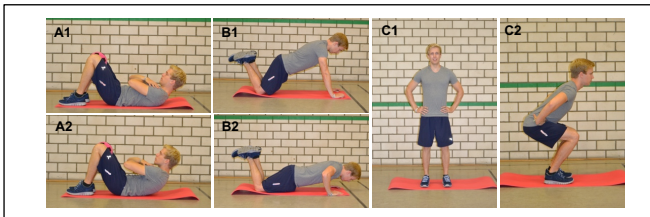


Figure 1: Exemplary strengthening exercises. A=sit-up; B=push-up; C= squat; 1=starting position; 2=end position.

Results

Children with PWS showed statistically significant increased local muscle strength after TI for single parameters (mean±standard deviation; pre / post: sit-ups (n): TI: 3.2±4.0 / 4.4±4.4, p=0.03 – CI:

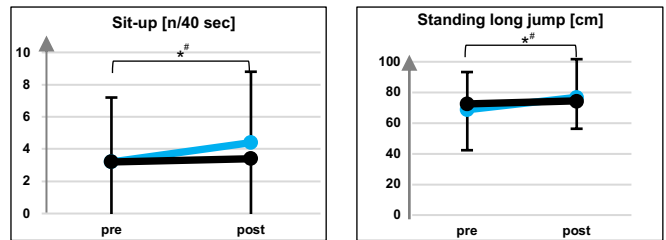


Figure 2: Local muscle strength of children with Prader-Willi Syndrome after a 10-week training intervention (TI) as well as a 10-week control intervention (CI). –TI –CI. *p≤0.05. Mean±standard deviation. #=significant alterations only for the training intervention.

3.2±3.4 / 3.4±3.6, p=0.93 and standing long jump (cm): TI: 68.7±26.4 / 76.8±24.9, p=0.02 – CI: 72.5±20.8 / 74.5±18.2, p=0.51) (Fig. 2). In contrast, no significant changes of any parameter could be observed following CI. Body weight of the children showed a downward trend after TI (mean±standard deviation; pre / post (kg): 58.7±8.9 / 56.9±7.9, p=0.06), but remained unchanged after CI (mean±standard deviation; pre / post (kg): 57.9±8.0 / 57.8±8.0, p=0.62). Furthermore, aerobic fitness and push-up performance tended to improve after TI, whereas the same parameters rather decreased after CI. In contrast to this, balance is tentatively improved after CI and unchanged after TI (Fig. 3).

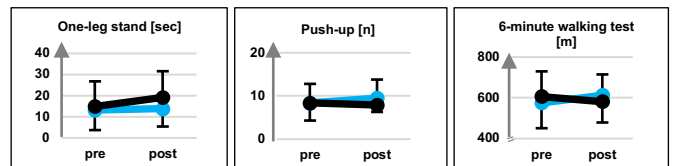


Figure 3: Balance, local muscle strength and aerobic fitness of children with Prader-Willi Syndrome after a 10-week training intervention (TI) as well as a 10-week control intervention (CI). –TI –CI. Mean±standard deviation.

Conclusion

This results demonstrate that a specific TI in form of a longitudinal home-based group training is feasible (in the setting of PWS living groups) and has positive effects on physical performance of children with PWS. Moreover, in order to increase physical performance of children with PWS, a specific training intervention seems more effective than an unstructured training intervention.

It is essential to integrate regular targeted physical training into a comprehensive treatment management and, therefore, into the daily life of children with PWS. For this, further studies have to develop more detailed recommendations regarding the intensity and duration of exercise interventions for children with PWS in order to improve their limited physical performance in an optimal manner.

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[3] Reusa L, van Vlimmerena LA, Bart Staal J, Ottenc BJ, Nijhuis-van der Sandena MWG. The effect of growth hormone treatment or physical training on motor performance in Prader-Willi syndrome: A systematic review. Neuroscience and Biobehavioral Reviews 2012; 36:1817–1838.