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Original Research Article

Relationship between physical activity and functional ability in school-aged children with hemophilia

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ABSTRACT

Introduction: With the advances in clotting factor replacement therapy children with hemophilia are increasingly encouraged to participate in physical activities and sports. Despite this positive trend, children with hemophilia still tend to be less physically fit than their healthy peers.
Aim: The main purpose of this study was to assess physical activity in school-aged children with hemophilia and its association with their functional ability, joint health and physical parameters.
Material and methods: Research material consisted of 24 boys aged 7–17 (mean 12.58±3.01 years) with severe or moderate hemophilia A or B. Weight, height and body mass index (BMI) were measured. Subjects activity level was assessed with Pediatric version of Hemophilia Activities List (PedHAL), joint health with Hemophilia Joint Health Score (HJHS version 2.1), functional ability with a Six-Minute Walk Test (6MWT).
Results and discussion: In Lithuanian children with hemophilia reduced physical activity (mean 83.64±11.40 scores) and functional ability (mean 408.46±68.58 m) were revealed. Strong negative correlation was found between PedHAL and HJHS scores ($r=-0.962$, $p<.0001$), HJHS and 6MWT ($r=-0.938$, $p<.0001$), strong correlation between PedHAL and 6MWT ($r=0.903$, $p<.0001$) scores. Distance walked displayed inverse correlation with age ($r=-0.858$, $p<.0001$), height ($r=-0.788$, $p<.0001$) and weight ($r=-0.894$, $p<.0001$).
Conclusions: Lithuanian children with hemophilia showed reduced physical activity and functional ability when compared with their healthy peers. The less joint impairments the subject had, the higher level of their physical activity and functional ability was. Age, height and weight were determinants of 6 minutes walking distance.

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1. Introduction

Motor fitness is considered an individual health measure. Its impact on human organism is extremely important during the early school years owing to the intensive growth at that time.[20] Although children are more agile than adults, they are at risk of consequences of hypokinesia due to the common forms of passive leisure activities.[19] Children with hemophilia are no exception to this notion. As a result of parental or medical restrictions, children with hemophilia often avoid any physical activity in their everyday life. Meanwhile, a physically active lifestyle is essential to maintain musculoskeletal health, reduce the risk of complications of hemophilia and ensure better quality of life in patients with this condition.[12,17,23,24] With the advances in clotting factor replacement therapy, children with hemophilia are increasingly encouraged to participate in physical activities and sports. In addition, it was noted that the attitude towards sports among patients with hemophilia has improved, and that the range of sports practiced has increased, presumably due to the improved medical treatment.[8,22] Despite this positive trend, children with hemophilia still tend to be less physically fit than their healthy peers. Koch et al. evaluated physical fitness of children with hemophilia aged 8.3–15.5 and reported a significant reduction in exercise capacity (peak work rate), possibly because of insufficient intensity of daily physical activities.[16] Engelbert et al. reported that children with hemophilia have a decreased aerobic capacity, lower reported leisure-time activity and less involvement in intense activity compared to their healthy peers.[5] Furthermore, Hassan et al. reported that children with hemophilia showed a decrease in distance achieved in the Six-Minute Walk Test (6MWT).[13] Muscle strength and anaerobic power were also significantly reduced in children with hemophilia, especially in the lower limbs.[14] In addition, children with hemophilia may be at increased risk of becoming overweight or obese as a result of inactivity because of joint bleedings or overprotection.[3] The need for a physically active lifestyle in patients with hemophilia is further highlighted by the finding that bone mineral density in children with severe hemophilia (FVIII/IX < 1%) is lower than in healthy subjects.[7]

2. Aim

The main purpose of this study was to assess physical activity in school-aged (7–17 years old) children with hemophilia and its association with functional ability, joint health and physical parameters.

3. Material and methods

This research was conducted in the Physical Medicine and Rehabilitation Center of Children's Hospital, Affiliate of Vilnius University Hospital Santariskiu Klinikos. Research material consisted of 24 boys aged 7–17 who suffered from severe or moderate hemophilia A or B. Weight and height were measured and a body mass index (BMI) was calculated (weight/height, kg/m^2).

Children's physical activity level was assessed using the Pediatric version of Hemophilia Activities List (PedHAL).[11] It contains 53 items across 7 domains and can be completed by both parents and children themselves. A raw score is converted to a normalized score that ranges from 0 (worst functional status) to 100 (best possible functional status).

Joint health of subjects was assessed with the use of the Hemophilia Joint Health Score (HJHS) version 2.1 by the International Prophylaxis Study Group (IPSG).[15] Joint function was measured with the HJHS, an eight-item scoring tool for the assessment of joint impairment of the six key index joints: knees, elbows and ankles. These eight items include duration of swelling, severity of swelling, muscle atrophy, crepitus on motion, flexion loss, extension loss, joint pain and muscle strength. Items are scored by grade of severity of impairment. The component joint scores were calculated, and an overall summarized score supplemented with a global gait score (i.e. observation of walking performance, stair climbing, running, and hopping on one leg). Thus, a final HJHS score ranges from 0 (no impairment) to 124 (maximum impairment for the six main joints).

Functional ability was determined by means of 6MWT.[1] It is a sub-maximal test of aerobic capacity, in which subjects walk as far as possible in 6 minutes around a pre-measured distance. It is a useful assessment tool for children with chronic conditions affecting the musculoskeletal system, because walking is a part of their everyday life.[13] In addition, heart rate was measured before and immediately after the 6MWT in a sitting position.

3.1. Statistics

Data analysis was performed using IBM SPSS Statistics 20.0 software. The results were analyzed and compared with the use of Student *t*-test and Pearson's correlation coefficient. Differences were considered statistically significant at $p < .05$.

4. Results

4.1. Subjects

Subject characteristics are presented in Table 1. Mean age of boys at the time of the study was 12.58 ± 3.01 years (range 7–17). A total number of children with severe hemophilia (FVIII/IX < 1%) was 21 and with mild hemophilia (FVIII/IX 6%–40%) – 3. Patients with severe hemophilia were treated prophylactically, whereas all patients with mild hemophilia received an 'on demand' treatment. All 24 subjects had joint impairments (mean 18.46 ± 7.28). In nearly half of the patients (45%) target joint was located at the ankle, in 33% at the knee, while the elbow was a target joint less frequently (21%).

4.2. Physical activity

All of the 24 PedHAL questionnaires were completed adequately and used for analysis. It took patients approximately 10 minutes to complete PedHAL questionnaire and there