Research in Developmental Disabilities 34 (2013) 4395–4403

Contents lists available at ScienceDirect

Research in Developmental Disabilities

Accelerometer-determined physical activity and walking capacity in persons with Down syndrome, Williams syndrome and Prader–Willi syndrome

Marianne Nordstrøm [a,e,*], Bjørge Herman Hansen [b], Benedicte Paus [c,d], Svein Olav Kolset [e]

[a] Frambu Resource Centre for Rare Disorders, Sandbakkveien 18, 1404 Siggerud, Norway
[b] Norwegian School of Sport Sciences, Department of Sports Medicine, Sognsveien 220, 0863 Oslo, Norway
[c] Oslo University Hospital, Department of Medical Genetics, P.O. Box 4950, Nydalen, 0424 Oslo, Norway
[d] University of Oslo, Institute of Clinical Medicine, Department of Medical Genetics, P.O. Box 1046, Blindern, 0317 Oslo, Norway
[e] University of Oslo, Institute of Basic Medical Sciences, Department of Nutrition, P.O. Box 1046, Blindern, 0317 Oslo, Norway

ARTICLE INFO

Article history:
Received 2 August 2013
Accepted 10 September 2013
Available online 18 October 2013

Keywords:
Accelerometer
Physical activity
Six-minute walk test
Down syndrome
Williams syndrome
Prader–Willi syndrome

ABSTRACT

In this study we describe by use of accelerometers the total physical activity (PA), intensity pattern and walking capacity in 87 persons age 16–45 years with Down syndrome (DS), Williams syndrome (WS) and Prader–Willi syndrome (PWS). Participants were recruited from all over Norway, and lived either with their parents or in community residences with support.

On average the participants generated 294 counts per minute (cpm) or 6712 steps per day, with most of the day spent in sedentary activity, 522 min/day, followed by 212 min/day in light PA, 71 min/day in lifestyle activity and 27 min/day in moderate-to-vigorous physical activity (MVPA). Inactivity was prevalent, as only 12% meet the current Nordic recommendations for PA.

When compared, no differences for total physical activity or time in MVPA were observed between the three groups. However, participant with DS spent a mean of 73 min/day less and 43 min/day less in sedentary activities compared to participants with PWS and WS, respectively, ($p = 0.011$, 95% CI: −10.9; −80.1). In addition the DS-group spent a mean of 66 min/day more in light PA than the PWS-group and 41 min/day more than the WS-group, ($p < 0.001$, 95% CI: 29.3; 79.7). Participants with PWS spent on average 30 min/day less in lifestyle activities compared to both participants with DS and WS, ($p < 0.001$, 95% CI: −14.2; −45.4). No association between total PA and BMI were observed. Males were more active than females across all diagnoses. Males accumulated on average 85 counts per minutes more than females, ($p = 0.002$, 95% CI: 33.3; 136.7), 2137 more steps per day, ($p = 0.002$, 95% CI: 778; 3496). The mean walking capacity during six-minutes was 507 m (SD 112 m) for males and 466 m (SD 88 m) for females. Distance walked during testing decreased with 33.6 m when comparing normal or underweight participants to overweight participants, and 78.1 m when comparing overweight to obese participants ($p < 0.001$ 95% CI: −40.4; −85.8). When adjusted for BMI no differences in walking capacity between the three genetic conditions were observed.

* Corresponding author. Tel.: +47 64856047; fax: +47 64856099.
E-mail addresses: mal@frambu.no, marianne.nordstrom@medisin.uio.no (M. Nordstrøm), b.h.hansen@nih.no (B.H. Hansen), benedicte.paus@medisin.uio.no (B. Paus), s.o.kolset@medisin.uio.no (S.O. Kolset).

0891-4222
http://dx.doi.org/10.1016/j.ridd.2013.09.021

1. Introduction

Regular physical activity (PA) and physical fitness improves functional ability, enhances independence and reduces the risk of non-communicable disorders such as cardiovascular disease, diabetes and several cancers (Nordic Council, 2005; WHO, 2010). The effectiveness of PA in relation to health depends on frequency, duration and intensity of activity, and is inherently difficult to assess due to its complex nature (Westerterp, 2009). Objective assessment of activity using activity monitors overcome many of the challenges related to self-reported measures of PA, such as social desirability bias and recall bias. Thus, accelerometers provide valid and reliable estimates of the degree, nature, and pattern of physical activity (Westerterp, 2009).

Walking capacity is a measurement for everyday physical capacity and cardiovascular fitness and is related to both levels of independence (Cowley et al., 2010) and long term health outcomes (Rasekaba, Lee, Naughton, Williams, & Holland, 2009). Walking is the most common form of exercise and PA in groups with intellectual disability (Draheim, Williams, & McCubbin, 2002). The six-minute walk test (6MWT) is a feasible and objective submaximal exercise test that assesses the distance a person can walk in six minutes (ATS Statement, 2002). In a variety of patient groups a distance less than 350 m walked in 6MWT is associated with increased mortality (Rasekaba et al., 2009).

Down syndrome (DS), Williams syndrome (WS) and Prader–Willis syndrome (PWS) are genetic conditions associated with mild or moderate intellectual disability. DS is caused by the presence of an extra copy or major portion of chromosome 21 (Hattori et al., 2000), WS by a deletion of the elastin gene on chromosome 7q11.23 (Morris, 2010) and PWS by the absence of paternally expressed genes in the 15q11-q13 chromosome region due to deletion, maternal disomy 15 (UPD) or an imprinting defect (Cassidy, Schwartz, Miller, & Driscoll, 2012). Even though molecular diagnosis is available today for all three conditions, diagnosis is in some patients based on clinical manifestations alone, especially in the adult patient population. Clinical manifestations of DS include typical physical, cognitive and behavioral characteristics which in most cases are easily recognized (Hunter, 2010). WS are recognized by typical facial features, short stature and connective tissue abnormalities in addition to a unique social and cognitive profile (Morris, 2010). Characteristics of PWS includes hypotonia, hypogonadism, a unique behavioral profile and childhood onset of hyperphagia that in absence of energy restriction will lead to obesity (Cassidy et al., 2012). It has previously been reported that individuals with DS and PWS have increased risk of inactivity and reduced physical capacity (Butler, Theodoro, Bittel, & Donnelly, 2007; Phillips & Holland, 2011; Temple & Stanish, 2009a), whereas specific knowledge on PA and everyday physical capacity in relation to WS is sparse.

In Norway all institutions for persons with intellectual disabilities closed, and individuals are offered supported living in community settings when moving from their parental homes (Beadle-Brown, Mansell, & Kozma, 2007). Independent living of individuals with intellectual disability in community settings has previously been associated with inactivity, low physical fitness and obesity (Doody & Doody, 2012; Draheim et al., 2002; Emerson, 2005; Hove, 2004; Martinez-Leal et al., 2011; Robertson et al., 2000). However, this knowledge is based on studies from countries with mixed types of living arrangements, where several confounding factors associated with persons living in institutions and persons living independently in communities, respectively, may occur. The complete deinstitutionalization in Norway opens an unique opportunity for studies of PA and physical capacity in a setting with increased focus on autonomy for all individuals, and for description of similarities and differences between subgroups associated with intellectual disability.

The aim of this study was (1) to describe levels of accelerometer-determined overall PA and sedentary behavior among persons with DS, WS and PWS; and (2) to investigate PA and walking capacity in relation to body mass index (BMI).

2. Methods

2.1. Ethical approval and recruitment procedures

Ethical approval for the study was granted by the Regional Committees for Medical and Health Research Ethics, South-East region.

Participants were recruited through existing information channels in relevant national-wide patient organizations, such as websites, membership bulletins etc. In addition a study-specific website was developed where general information, formal letter-to-participate and consent to participate-scheme was posted and available to download. In order to be eligible for inclusion, the individuals had to be between 16 and 45 years of age, and diagnosed with either DS, WS or PWS verified by standardized clinical methods (Holm et al., 1993; Preus, 1984) or by laboratory genetic testing. We used a convenient sampling frame. All participants who returned written informed consent to participate-scheme, signed by both the participant and legal guardian/parent were invited to participate.

2.2. Participants

A total of 96 participants, 40 with DS, 28 with WS and 28 with PWS from all over Norway participated in the study. All participants with clinical diagnosis were offered voluntary genetic testing for verification of their condition. Nine participants were eliminated from final analysis due to negative result from laboratory genetic testing and not for filling clinical criteria. In total 27 participants with DS have genetically verified trisomy 21 and 13 have clinically diagnosis. Of Participants with WS 18 have a genetically verified diagnosis whereas 7 have a clinically diagnosis (Preus, 1984). In the