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Research Article

Motor proficiency in young children with Prader-Willi syndrome: a preliminary report

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HIGHLIGHTS

- This study used a norm-based test to document motor proficiency in children ages 4-6 with Prader-Willi syndrome (PWS).
- This is the first study to use all items in the Bruininks-Oseretsky Test of Motor Proficiency-2 (BOT-2) short form in young children with PWS.
- This study showed that young children with PWS who had been on growth hormone replacement for three or more years still exhibited well-below average motor proficiency.
- Multiple subtest items in the BOT-2 short form exhibited floor effects questioning the suitability of this tool in young children with PWS.

ABBREVIATIONS

BOT-2-SF	Bruininks-Oseretsky Test of Motor Proficiency-Second Edition
GH	Growth hormone
GHRT	Growth Hormone Replacement Therapy
MP	Motor proficiency
NT	Neurotypical development
PWS	Prader-Willi syndrome

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BACKGROUND: Systematic documentation of motor characteristics in young children with Prader-Willi syndrome (PWS) is vital as access to treatments improves.
AIM: To characterize motor proficiency (MP) in young children with PWS.
METHOD: Participants included 6 children (3 male and 3 female) with PWS and 13 children with neurotypical development (NT), (9 male and 4 female) ages 4-6 years. Five out of six children with PWS had been on growth hormone replacement therapy (GHRT) for >3 years. Some children with PWS exhibited cognitive delays and others performed within the average range (Intellectual quotient mean± standard deviation = 65.3 ± 7.62, range = 47 – 94). MP was measured using the Short Form of the Bruininks-Oseretsky Test of Motor Proficiency-Second Edition (BOT-2-SF).
RESULTS: Children with PWS scored lower than children with NT in all areas of MP except for fine motor integration. All children with PWS scored well-below average for total MP; children with NT scored average (n=10) or above-average (n=3) for total MP, respectively.
CONCLUSION: At this young age children with PWS universally exhibited poor MP despite most of them being on GHRT and some exhibited intellectual functioning in the average range. Evidence of BOT-2-SF floor effects underscores the need to refine assessment procedures and enhance measurement precision for this population.

KEYWORDS: Motor skills | Neurodevelopment | Obesity | Genetics | Prader-Willi syndrome

INTRODUCTION

Prader-Willi syndrome (PWS) is a rare neurodevelopmental disorder occurring in approximately 1 in 15,000 individuals [1]. PWS results from genetic changes to the q11.2-q13 region in the paternal chromosome 15. The three genetic changes that cause PWS include a deletion in the genes, an imprinting-center defect, or the expression of two maternal chromosomes 15 (called uniparental disomy) but no paternal chromosome 15. The main alteration in the genes causes hyperphagia that sets on during childhood and continues onto adulthood[1].Individuals with PWS also present with hypotonia, low lean mass, excessive body fat, growth hormone (GH) deficiency, cognitive delays, and behavioural challenges [1]. Hyperphagia is managed with behavioural strategies, a caloric-restricted diet and physical activity to prevent unhealthy weight gain [1,2].

Anecdotal evidence indicates infants and toddlers with PWS reach their developmental motor milestones at twice the age as their peers with neurotypical development (NT)[1,3]. This delayed achievement of motor milestones appears related to muscular hypotonia, poor muscle mass, GH deficiency (due to effects on muscle and neuroplasticity), and intellectual disability [3,4]. The use of recombinant GH is nowadays standard of care in PWS [1]. Studies with recombinant GH in infants demonstrate improvement in lean mass and gross motor development [5,6] and cognitive development [4]. Studies that have followed infants into childhood treated with recombinant GH show

improvements in lean mass, decreased fatness and improved cognitive functioning compared to untreated controls with PWS [7]. In children with NT ages 1 month to 4 years, physical activity interventions are associated with improved motor and cognitive development [8].

Motor proficiency involves competence or mastery in fine and gross motor skills involving motor precision, fine and gross motor coordination, balance, muscular strength, agility and movement speed. In children with NT, early motor development is related to later motor and cognitive development, which will affect acquisition and mastery of skills [9]. Obesity has been negatively associated with motor skills [10] while physical activity has been positively associated with motor skills [11,12].

In older children with PWS, hypoactivity, increased adiposity, decreased lean mass, GH deficiency, and delays in cognitive functioning are among the factors speculated to negatively influence motor proficiency during childhood [6,13]. In contrast, recombinant GH therapy improves aspects of motor proficiency [6]. And, participation in skill-building physical activity improves motor proficiency in children with and without PWS [14]. As nowadays, many children with PWS have received recombinant GH since infancy, that has changed their natural history [15]. Thus, characterization of motor proficiency in young children with PWS is important to help guide early age therapeutic interventions with the aim of increasing PA. Therefore, the purpose of this preliminary study was to compare motor proficiency in young children with PWS to an age-matched group of children with NT using a previously validated norm-based test of motor proficiency.

METHODS

Participants

Participants were recruited through the Prader-Willi California Foundation by contacting potential families with a child with PWS ages 4-7 years by mail or at events. Participants with NT ages 4-7 were recruited through health fairs and word of mouth. Seventeen families with a child with PWS indicated interest to participate in the study. Four families scheduled to enroll in the study but did not show at the time of the visit. Six more families with a child with PWS also indicated interest but did not participate. Seven participants with PWS were enrolled; however, one participant was withdrawn because could not participate in the activities because had difficulties understanding the task instructions. Thirty-three families with a child with NT indicated interest and 20 enrolled. However, to match the age between groups, only participants with NT ages 4-6 were included in this study.

Participants with PWS were 6 children (3 male and 3 female) aged 4-6 years and 13 children with NT (9 male and 4 female) aged 4-6 years. Children with PWS diagnoses included: 2 maternal uniparental disomy, 2 maternal uniparental disomy or imprinting defect, 1 imprinting defect, and 1 deletion. Five children with PWS were receiving growth hormone replacement therapy (GHRT). Children with PWS were currently participating in physical therapy (83.3%), and occupational therapy (83.3%). The children with NT had no known history or diagnosis of developmental delays or neurodevelopmental disorders. Verbal assent was obtained from child participants and written consent forms were signed by their parent before participation. This study was approved by the California State University Fullerton (CSUF) Institutional Review Board protocol HSR#17-18-389 and the United States Army Human Protection Research Office and followed the 1975 Declaration of Helsinki.

Overall Procedures

Participants' parents completed a medical and exercise history questionnaire to screen for potential health-related risks to exercise. Parents (N =18, PWS=5, NT=13). reported the approximate age in months in which their children achieved these developmental motor milestones: stood alone well, walked alone, ran alone without falling, and manipulated object purposefully. Children were measured for intellectual functioning, anthropometrics, and motor proficiency.

Measurements

Intellectual Functioning

Intellectual functioning was assessed by a licensed clinical psychologist with a specialization in neurodevelopmental disorders. Child IQ was estimated using the Abbreviated Battery Intelligence Quotient from the Stanford-Binet Intelligence Scales, Fifth Edition [16], which is comprised of two subscales with high loading on the general intelligence factor: a non-verbal fluid reasoning task and a verbal knowledge test. The abbreviated battery intelligence quotient scores have a mean of 100 and a standard deviation of 15.

Anthropometrics

Participants were measured for height without shoes using a wall-mounted measuring tape and at the end of inhalation. Body mass was measured with no shoes in shorts and t-shirt using a digital scale (The Health-O-Meter Professional, Model 5i99KL PelStar, Alsip, IL USA). Body mass index percentile was calculated.

Motor Proficiency

Children's MP was measured using the Bruininks-Oseretsky Test of Motor Proficiency-Second Edition Short Form (BOT-2-SF) [17]. The BOT-2-SF includes 4 motor-area composites: fine manual control, manual coordination, body coordination, and strength and agility. Fine manual control includes subtests for fine motor precision (2 test items) and fine motor integration (2 test items). Manual coordination includes subtests for manual dexterity (1 test item) and upper-limb coordination (2 test items). Body coordination includes subtests for bilateral coordination (2 test items) and balance (2 test items). Strength and agility include subtests for running speed and agility (1 test item) and strength (2 test items). The BOT-2-SF provides a total point score from all subtests, converted into an age and sex standard score. Standard scores for the BOT-2-SF range from 20 to 80 for ages 4 to 7 with a score of 50 placing a child's performance in the 50th percentile rank for age and sex. From this standard score, performance is classified into descriptive categories: well-below average, below average, average, above average, and well-above average. For individual subtests, only point scores derived from raw scores (non-standardized by sex or age) are obtained. The BOT-2 has shown good to excellent test-retest reliability (ICCs 0.74-0.97) for the composite score and most motor areas, with fair-to-poor reliability for bilateral coordination (ICC 0.60), strength (ICC=0.57) and running speed and agility (ICC=0.47) in youth with PWS ages 8-11 years [18]. In addition to the strength subtests of push-ups and sit-ups, children completed the long jump and wall sit subtests from the long form of the BOT-2. These additional subtests were included to obtain modified scores for strength and overall motor proficiency given evidence of considerable difficulty with the push-up and sit-up subtests in pilot testing with a separate preschool-aged sample of children with PWS (n=3).

Two experienced examiners conducted the assessments. Testing occurred in the Movement Enhancement Laboratory at CSUF, a climate-controlled (~20° C) space with a large open area free of distractions. All children with PWS were assessed during the morning to avoid the afternoon lethargy. Children without PWS were assessed in the morning or early afternoon based on convenience of the families.

Data Processing and Analyses

Independent sample Mann-Whitney U-tests were used to compare groups based on age, physical characteristics, parent-reported major developmental motor milestones achievement, motor proficiency subscales, and total motor proficiency. The non-parametric Mann-Whitney U-test was selected given the small sample size of children with PWS and the unequal sample size between groups. For the BOT-2-SF, frequencies were calculated for percent of participants in each group presenting with well-below average, below average, average, above average, and well-above average motor proficiency performance. Statistical significance was set at $p < .050$.

RESULTS

There were no differences for age, height, body mass or body mass index percentile between children with PWS and children with NT. Children with PWS exhibited delays in intellectual functioning. However, performance varied considerably, with some children with PWS demonstrating intellectual functioning within the average range (Intelligence quotient *Mean* = 65.3, *SD* = 7.62, Range = 47 - 94). Children with PWS reached the major developmental motor milestones (i.e., stood alone, walked alone, ran alone and manipulated object purposefully) at a later age than children with NT (See Table 1).

Children with PWS scored lower than children with NT in the overall motor proficiency score and in all individual areas except for fine motor integration (See Table 2). All children with PWS scored in the well-below average descriptive category for motor proficiency, while all children with NT scored either in the average (*n* = 10, 76.9%) or above-average (*n* = 3, 23.1%) categories when considering the standard administration of the BOT-2-SF. In the modified BOT-2-SF those with NT scored in the average (*n* = 9, 69.2%) and well-above average range (*n* = 4, 30.8%). For those with PWS, there were no differences when the BOT-2-SF was modified.

Table 1. Participant characteristics of children with PWS and children with NT presented as median (minimum-maximum).

	Children with PWS (*n* = 6)	**Children with NT (*n* = 13)**
Male/Female	3/3	9/4
Age (years)	5 (4-6)	5 (4-6)
Height (cm)	108.6 (99.1-119.4)	113.0 (99.1-132.7)
Body mass (kg)	21.9 (15.0-33.5)	21.3 (16.4-43.5)
Body mass index percentile	91 (51-99)	61 (17-99)
Motor Milestones[a]	(*n* = 5-6)	(*n* = 10-13)
Stood alone (months)	24 (21-28)*	11 (8-18)
Walked alone (months)	25.5 (17-39)*	12 (10-18)
Ran alone (months)	41 (30-54)*	14 (11-23)
Manipulated object purposefully (months)	24 (13-36)*	8.5 (5-13)

PWS, Prader-Willi syndrome; NT, Neurotypical development
* = Different than children with NT, $p < 0.001$
[a] Not all parents could recall the time of achievement of all motor milestones so number of participants for each milestone varied.

Table 2. Results of the Bruininks-Oseretsky Test of Motor Proficiency-2 Short Form in children with PWS and children with NT of similar chronological age (ages 4-6 years).

	Children with PWS (*n* = 6)	**Children with NT (*n* = 13)**	
	median (range)	**median (range)**	***p*-value**
Fine motor precision	1.5 (0-5)	7 (0-10)	0.009
Fine motor integration	2 (0-5)	5 (0-9)	0.072
Manual dexterity	2 (0-3)	5 (2-6)	<0.001
Upper-limb coordination	0 (0-2)	6 (0-11)	0.002
Bilateral coordination	0 (0-0)	6 (0-7)	<0.001
Balance	1.5 (0-3)	6 (3-8)	<0.001
Speed and agility	0 (0-0)	8 (1-9)	<0.001
Strength	0.5 (0-3)	5 (0-8)	0.009
Strength modified	1.5 (0-3)	6 (2-9)	<0.001
Total motor proficiency (standard score)	25.2 ± 3.1 (21-30)	53 (44-67)	<0.001
Total motor proficiency modified (standard score)	25.0 ± 3.8 (21-31)	55 (48-68)	<0.001

Notes: All area scores are presented in point scores, and total motor proficiency scores (20-80 range) are presented in standard scores (standardized to age and sex); Fine motor integration children with NT (*n* = 11), Strength modified and Total motor proficiency modified scores included subtest items of Standing Long Lump and Wall Sit.

DISCUSSION

These preliminary results demonstrate poorer motor proficiency in young children with PWS aged 4-6 years than age-matched peers with NT based upon standardized, norm-referenced score interpretations. These findings expand upon the previously documented low levels of motor proficiency shown in children with PWS ages 8-11 years [13]. These results suggest that efforts to foster the achievement of motor proficiency in those with PWS are of utmost importance in early childhood.

Motion is limited in infants with PWS due to severe hypotonia and muscle weakness [1], and this may delay gross motor development [6]. In the present study children with PWS reached major developmental motor milestones at twice the time of the subsample with NT, consistent with anecdotical reporting [3]. And, before reaching the milestone of standing alone well four children with PWS had been on GHRT 4-19 months. The use of GHRT leads to improvements in lean mass at early ages, and consequently motor performance [5,19]. However, in the present study five out of six children had been on GHRT for more than two years at the time of this study. This fact highlights that while GHRT improves motor proficiency and could be helpful in enhancing rehabilitation and training [19] other factors influence motor proficiency at this young age and routines should be implemented for achievement of gross motor skills.

In one of the running speed and agility subtests, children were asked to hop on one leg, a milestone achieved at 3-5 years of age in children with NT [20]. None of the children with PWS were able to hop and leave the ground indicating deficits in muscular force. Children with PWS were able to complete the long jump subtest but still scored lower than the controls with NT. Jumping performance relies on the ability of the muscles in the lower limbs to produce power during the push-off phase [21]. In could be argued that hopping also demands force and speed. In young children, short-term muscular power needed to jump or hop depends on muscular volume which depends on muscle mass [22]. However, neurological factors such as motor unit activation and synchronization, while not completely understood because of technical limitations, cannot be overlooked [23]. Specifically, improvement in muscular strength with resistance training in children with and without PWS can occur independently of changes in lean mass [14,23]. Additionally, both tasks also demand coordination of the different body segments and inability to coordinate may result in inefficiency [23]. While children with NT show lower ability to recruit and fully activate these high threshold motor units compared to adults [24], adults with PWS still exhibit a decreased ability to activate these motor units [25]. Hence, young children with PWS may exhibit difficulty with these tasks due to multiple aspects related to force, power, and coordination. Professionals working with young children with PWS may utilize adapted jumping exercises (such as jumping down) or use therapies like whole-body vibration platforms to stimulate neuromuscular activation [26]. Conversely, the smaller difference in scores related to motor integration between those with PWS and those with NT also show how fine motor skills that are routinely practiced (such as copying figures and patterns) and do not rely on muscle force show less difference in performance.

The intellectual functioning of children with PWS in the present study was variable, with some children displaying significant cognitive delays and others performing within the average range. Reduced cortical complexity in children with PWS has been related to cognitive and developmental delays [27], and lower levels of intellectual functioning have been associated with poorer MP as evidenced in fine and gross motor skills [28]. In the present study, it was sometimes challenging to differentiate task comprehension difficulties from difficulties tied more directly to motor proficiency. In our study, children with PWS struggled significantly on a multi-component test of bilateral coordination (alternating feet and tapping hands), with all scoring zero on this subtest.

Indeed, the BOT-2-SF, may have been too long for those with PWS (approximately 40-60 minutes) compared to the suggested 15-20 minutes. Participants required more and longer breaks in between subtests and repeated instructions. Some subtests, particularly those requiring attention to detail and comprehension and enactment of complex, multi-component instructions, may have been too difficult to complete, resulting in floor effects. For instance, to earn full points on the balance beam test, children must, for ten seconds, balance on one foot, keep both hands placed on the hips, and have the opposite leg bent at 90 degrees. Many children with PWS were able stand on the beam on one foot, but often could not meet all of the requirements earning a score of zero. Another test item from BOT-2 (walking forward on a line heel-to-toe), has also been identified as not being feasible even for young children with NT [29]. Hence, performance on balance tasks as well as on some other tasks mentioned previously raise the question as to the suitability of the BOT-2-SF for use with this young age-group, particularly for those with PWS.

As some of the issues experienced by participants with PWS could be experienced in other neurodevelopmental disorders, future studies could benefit from findings of this study regarding limitations of the BOT-2-SF. We substituted the strength subtests of push-ups and sit-ups with a long jump and wall sit (also from the BOT-2). While this allowed the children to complete the tasks these substitutions did not result in any differences for the composite score. Perhaps, other substitutions could be explored and validated such as the use of a wall push-up in addition to other measurements for speed and agility.

Our study was not without limitations and our sample size was small. Recruitment of children aged 4-6 years with PWS was rather difficult because of the competing therapies that children this age receive (e.g., speech, behaviour, and physical therapy to name a few), which can make participation in research studies outside of clinics or hospitals more difficult. While we substituted some test items from the BOT-2 -SF with items from the long form we did not validate them. Bias in recalling motor milestone achievement by the parents should also be acknowledged.

CONCLUSION

These preliminary results showed that young children with PWS perform poorly in tests of motor proficiency. Additionally, floor effects due to difficulty of the test items suggest the need for selecting or developing and validating tests that may be more suitable to assess motor proficiency in children with neurodevelopmental delays and/or motor impairments similar to those exhibited by children with PWS.

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