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Examination of sensory reception and integration abilities in children with and without Prader-Willi syndrome

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Highlights

- This study used the Sensory Organization Test® to assess postural control in children with and without Prader-Willi syndrome.
- Children with PWS showed deficits in reception and processing of sensory information.
- Balance activities designed to improve sensory reception and integration skills in PWS are highly recommended.

Abstract

Background

Good postural stability control is dependent upon the complex integration of incoming sensory information (visual, somatosensory, vestibular) with neuromotor responses that are constructed in advance of a voluntary action or in response to an unexpected perturbation.

Aims

To examine whether differences exist in how sensory inputs are used to control standing balance in children with and without Prader-Willi syndrome (PWS).

Methods and Procedures

In this cross-sectional study, 18 children with PWS and 51 children categorized as obese but without PWS (without PWS) ages 8–11 completed the Sensory Organization Test®. This test measures the relative contributions of vision, somatosensory, and vestibular inputs to the control of standing balance. The composite equilibrium score (CES) derived from performance in all sensory conditions, in addition to equilibrium scores (EQs) and falls per condition were compared between groups.

Outcomes and Results

The CES was lower for children with PWS compared to children without PWS (M=53.93, SD=14.56 vs. M=66.17, SD=9.89, $p = .001$) while EQs declined in both groups between conditions 1 and 4 (F (1.305, 66.577) = 71.381, $p < .001$). No group differences in the percent of falls were evident in condition 5 but more children with PWS fell in condition 6 (χ^2 (1) = 7.468, $p = .006$). Group differences in frequency of repeated falls also approached significance in conditions 5 (χ^2 (3) = 4.630, $p = .099$) and 6 (χ^2 (3) = 5.167, $p = .076$).

Conclusions and Implications

Children with PWS demonstrated a lower overall level of postural control and increased sway when compared to children with obesity. Both the higher incidence and repeated nature of falls in children with PWS in conditions 5 and 6 suggest an inability to adapt to sensory conditions in which vestibular input must be prioritized. Postural control training programs in this population should include activities that improve their ability to appropriately weight sensory information in changing sensory environments, with a particular focus on the vestibular system.

What does this study add?

This study shows that children with PWS demonstrate a lower level of postural stability. The results suggest that children with PWS show inability to adapt to sensory conditions that require prioritizing vestibular information to maintain postural control. This information can be used to help guide training programs in this population.

Keywords

Sensory integration
Postural control
Obesity
Neurodevelopmental disorder

1. Introduction

Prader-Willi syndrome (PWS) is a rare genetic disorder characterized by early onset morbid childhood obesity, compulsive overeating, behavioral and cognitive impairments, and less overall spontaneous physical activity (Butler et al., 2007, Cassidy et al., 2012, Castner et al.,

2014, van den Berg-Emons et al., 2008, van Mil et al., 2000). Individuals with PWS have an abnormal body composition, specifically increased body fat, decreased lean mass, and hypotonia (Eiholzer & Whitman, 2004). Additionally, PWS presents with growth hormone deficiency, poor stamina, and lethargy (Holm et al., 1993; Reus et al., 2011).

Obesity, both congenital and non-congenital in origin, has been associated with poor balance and coordination. Individuals with obesity, with and without PWS, exhibit decreased movement skill competency including poor balance and coordination skills (Lam et al., 2016), potentially leading to less participation in physical activity (Elmesmari et al., 2018). Lower physical activity levels lead to lower muscular activation and strength and ultimately, poorer balance capacity (Capodaglio et al., 2011b). Children with PWS appear to experience even greater balance and coordination deficits attributed to hypotonia, as well as sensory integration deficiencies due to underdeveloped somatosensory and vestibular systems (Agarwal, 2009, Cataletto et al., 2011). As poor balance and weak muscles, among other factors, are associated with an increased risk of falls in older adults and persons with disability (Alghwiri & Whitney, 2019; Shi et al., 2015), this topic is of great concern in PWS due to the increased fracture risk previously documented in children (Kroonen et al., 2006) and adults with PWS (Butler et al., 2002).

The Sensory Organization Test (SOT)® is considered to be the "gold standard" for examining sensory impairments in the visual, somatosensory, and vestibular systems in relation to balance and has previously been used in pediatric populations with and without disabilities (Cherng et al., 1999, Westcott et al., 1997). Within the age range of 7 to 10 years, typically-developing children have demonstrated postural responses that have reached adult-like maturity, as well as an ability to resolve sensory conflict and appropriately weight the vestibular system when non-veridical information is available from the visual and somatosensory systems (Westcott et al., 1997). However, studies using the SOT® have demonstrated that postural stability and overall balance performance are reduced in conditions such as autism spectrum disorder (ASD) (Minshew et al., 2004), attention deficit hyperactivity disorder (Shum & Pang, 2009), cerebral palsy (Cherng et al., 1999), and developmental coordination disorder (DCD) (Fong et al., 2013).

To date, little attention has been focused on quantifying how children with PWS receive and integrate sensory information in altered sensory environments. Many studies have investigated postural stability and control in adults with PWS using static force plates (Capodaglio et al., 2011b, Cimolin et al., 2011, Cimolin et al., 2011, Galli et al., 2011); and dynamic force plate technology (Rubin et al., 2023). In particular, no studies have examined sensory reception and integration abilities in children with PWS to determine potential deficits affecting postural control unrelated to excess body fat. Therefore, the purpose of this study was to assess sensory reception and integration skills in children with PWS compared to children without PWS, but with obesity.

2. Material and Methods

2.1. Participants

Eighteen children with PWS and 51 children with obesity (without PWS) and ranging in age from eight to 11 years participated in this study (see Table 1). Obesity was defined as having a body fat percentage greater than or equal to the 95th percentile for age and sex (McCarthy et al., 2006). All participants with PWS provided genetic testing documentation to confirm diagnosis: deletion (n = 7), uniparental disomy (n = 3), uniparental disomy or imprinting defect (n = 3), or DNA methylation (n = 5). Children with PWS also reported currently (n = 15), previously (n = 1),

or never (n = 2) taking growth hormone replacement therapy. Children with PWS were currently engaged in physical therapy (n = 5), aquatic therapy (n = 3), horse therapy (n = 5) and structured physical activity (n = 10). Only two children without PWS had symptoms of dizziness related to exercise as indicated in the medical history form completed by parents. Three participants with PWS also reported a history of low body temperature, with one of the three also reported a history of high body temperature in their medical history form. Exclusion criteria included confirmed pregnancy, current usage of lipid-lowering diabetes, or blood pressure medications, or an inability to participate in moderate to vigorous physical activity. This study was approved by the Institutional Review Board at California State University Fullerton and the Human Subjects Research Protection Office from the United States Army Research and Materiel Command . Written informed assent and consent were obtained from all participants and their parents prior to participation.

Table 1. Demographic characteristics of children with and without PWS.

Empty Cell	**Children with PWS (*n* = 18)**	**Children without PWS (*n* = 51)**	**Total (*n* = 69)**
Empty Cell	**Mean (*SD*)**	**Mean (*SD*)**	**Mean (*SD*)**
Age	9.11 (1.13)	9.86 (1.13)	9.7 (1.2)
Height (cm)	137.93 (9.57)	146.26 (8.98)	144.1 (9.8)
Weight (kg)	49.61 (17.30)	60.15 (17.21)	57.4 (17.7)
BMI (kg/m^2)	25.87 (8.34)	27.68 (5.28)	27.2 (6.2)
Body Fat (%)	45.98 (9.95)	43.98 (5.79)	44.5 (7.1)
Empty Cell	*n* (%)	*n* (%)	*n* (%)
Sex			
Male	10 (55.6%)	25 (49.0%)	35 (50.7%)
Female	8 (44.4%)	26 (51.0%)	34 (49.3%)

2.2. Test Administrators

Four test administrators (AMT, EB, DJR, LS) conducted the SOT® tests. One of the co-principal investigators (DJR) with several years of experience administering this test served as the lead test administrator and trained three graduate student research assistants to administer the test over a three-week period. In addition to being trained on the protocol, the research assistants observed the lead test administrator deliver the test to at least two children with PWS prior to administering the test while being supervised. The lead test administrator also conducted fidelity checks throughout the study. The research assistants were already experienced with administering other physical performance tests to children with and without PWS and followed a written script while administering the test.

2.3. Procedures

Anthropometrics were measured in the children while wearing light clothing and no shoes using standardized procedures (Centers for Disease Control, 2007). Body mass was measured via calibrated electronic scale (ES200L, Ohaus, Pinewood, NJ) and stature via wall-mounted stadiometer (Seca, Ontario, CA). Body composition was measured using dual-energy x-ray

absorptiometry (DXA) following standard procedures (GE Healthcare, GE Lunar Corp., Madison, WI).

2.4. Sensory reception and integration skills

All participants completed the SOT® protocol on the SMART Balance Master® (NeuroCom International, Clackamas, OR) as described in the test manual. The SOT® consists of six testing conditions in which the visual surround and/or support surface are systematically manipulated. The programmed movement of the force-plate and/or visual surround during the SOT® forces the individual to reweight the sensory input in order to maintain standing balance. In certain conditions, visual input is eliminated while in other conditions the visual surround and/or support surface is sway-referenced (i.e., the anterior-posterior movement of the visual surround and/or support surface is directly proportional to the amount of sway of the individual) in order to render visual and/or somatosensory input inaccurate (Nashner, 1993). Specifically, in condition 1, all three sensory systems are available (i.e., no sensory environment manipulation); in condition 2, vision is removed (participant wears a blindfold) while standing on a stable support surface; in condition 3, the three-sided visual surround is sway-referenced, disrupting the use of vision to control upright balance, while the support surface below is stable; in condition 4, the eyes are open but now the support surface is sway-referenced; in condition 5, vision is again removed (participant wears a blindfold) and the support surface is sway-referenced; finally, in condition 6, both the visual surround and support surface are sway-referenced, creating a situation of sensory conflict between the visual and somatosensory systems and the need to prioritize vestibular inputs to control upright posture (see Table 2).

Table 2. The Sensory Environment and Available Sensory Input Across the Six Conditions of the Sensory Organization Test®.

Condition	Sensory Environment	Available Sensory Input
1	Normal vision; stable support surface	Vision; somatosensory; vestibular
2	Absent vision; stable platform	Somatosensory; vestibular
3	Sway-referenced vision; stable platform	Somatosensory; vestibular
4	Normal vision; sway-referenced platform	Vision; vestibular
5	Absent vision; sway-referenced platform	Vestibular
6	Sway-referenced vision and platform	Vestibular

Participants wore light clothing and no shoes or socks during testing. Before the test, each participant was fitted for an overhead safety harness to prevent injury from potential falls. Participants stood on the force plate of the SMART Balance Master® with the arms fully extended at the sides of the body. The feet were positioned in accordance with the manufacturer's requirements for data collection. Participants completed a familiarization trial in which the test was first verbally explained and then one trial of each of the six testing conditions was administered. A familiarization trial was considered necessary to lower anxiety levels in the children being tested and/or ensure that they understood and were able to follow the instructions for standing quietly during each sensory condition. After the familiarization trial, the child was given a three-minute rest in a seated position. The participant's feet were then repositioned on the force plate prior to the start of the test. Each participant completed three, 20-second standing trials in each of the six test conditions during a single test session of approximately 30 min. A mean Composite Equilibrium Score (CES) across all six test

conditions, condition-specific Equilibrium Scores (EQs), and the number of falls (defined as a loss of balance requiring external support from the overhead harness, visual surround, and/or the test administrator) in each condition were generated by the SMART Balance Master®.

2.5. Statistical analysis

Based on test administrator written comments following completion of each test condition, participants were excluded from further analyses if their data were deemed to be invalid. Participants were excluded if they were unable to maintain a quiet standing position during each test condition or appeared to "play" or "test" the equipment when either the visual surround and/or force plate were sway-referenced. A comparison between the performance of those children subsequently excluded and those included was completed. Preliminary analyses were also conducted to evaluate the data and assumptions for statistical tests using the mean CES and EQs. This included assessing normality of the data. Sphericity and the assumption of homogeneity of variance were evaluated when the individual statistical analyses were performed.

To assess differences between children with and without PWS, three distinct analyses were performed. First, an independent t-test was performed to identify group differences in the mean CES. Second, a 2 (group) by 4 (condition) factorial ANOVA was performed on the EQs for the first four sensory conditions. Only those children (with PWS = 11; without PWS= 42) who completed all three trials in each of the first four sensory conditions without registering a fall were included in this analysis. As the number of falls experienced by participants were higher in conditions 5 and 6, EQ scores for both conditions were excluded from the factorial ANOVA and a third analysis comparing the percentage of falls recorded for both groups in conditions 5 and 6 was conducted. Frequencies of falls within conditions 5 and 6 as a function of group (with PWS versus without PWS) were compared using chi-square analysis. In addition, children experiencing repeated falls (fell on trial 1 and 2; 1 and 3; or 1, 2, and 3) in either condition were compared to those who only fell on the first trial. Significance level for all statistical analyses was set at $p < .050$. IBM SPSS Statistics 23.0 for Windows (SPSS Inc., Chicago, IL) was used for the statistical analyses.

3. Results

3.1. Preliminary analysis

Four children with PWS (22.2%) and five children without PWS (9.8%) had invalid data on at least one of the trials in one or more of the six test conditions. These nine children were excluded from further analysis. No significant performance differences were observed between those with and without PWS in terms of valid data (χ^2 (1) = 1.809, $p = .179$). There were also no significant differences as a function of age or test administrator for children with and without PWS ($p > .050$). However, all five children without PWS who had invalid data were male ($p = .016$). For children with PWS, there was no difference in exclusion based on male and female with two males and two females excluded ($p > .050$).

Descriptive statistics including means, standard deviations, as well as skewness and kurtosis scores were calculated for both the mean CES and the individual EQs. In terms of normality, the CES as well as the majority of individual EQs showed standardized skewness and kurtosis values that were within acceptable ranges ($-3.29 < z < 3.29$), with the exception of the EQs for condition 3. This variable showed violations for both skewness and kurtosis.

3.2. Differences in equilibrium scores

The mean CES was lower for children with PWS (M=53.93, SD=14.56) when compared to children without PWS across conditions 1 through 4 (M=66.17, SD=9.89), $t(58) = 3.611$, $p = .001$, Cohen's d= 0.84. Due to normality being violated in condition 3, a square root transformation was performed on the EQs for conditions 1 through 4. The analysis was subsequently performed with and without transformed data. As the results were similar, the non-transformed data are reported for ease of interpretation. As a result of sphericity also being violated based on Mauchly's test, the Greenhouse-Geisser correction was used for the factorial ANOVA. Results from the 2 (group) x 4 (condition) factorial ANOVA revealed no group by condition interaction, $F(1.305, 66.577)= 1.433$, $p = .242$. However, the main effect for condition was significant, $F(1.305, 66.577)= 71.381$, $p < .001$. Post-hoc analysis using a Bonferroni correction revealed that performance across both groups was significantly different for conditions 2, 3, and 4 when compared to condition 1 ($p < .001$). No significant difference in EQs was evident between conditions 2 and 3 ($p = .451$). No main effect for group was evident, $F(1, 51)= 1.309$, $p = .258$ (see Table 3).

Table 3. Differences in Condition-specific Equilibrium Scores for children with (n = 11) and without PWS (n = 42).

Condition	Participants	Frequency	Equilibrium Score mean (SD)
1	All children	N = 53	89.80 (0.49)*
Empty Cell	With PWS	N = 11	89.33 (0.86)
Empty Cell	Without PWS	N = 42	90.27 (0.44)
2	All children	N = 53	87.11 (0.73)*
Empty Cell	With PWS	N = 11	87.06 (1.29)
Empty Cell	Without PWS	N = 42	87.16 (0.66)
3	All children	N = 53	85.98 (0.87)*
Empty Cell	With PWS	N = 11	85.49 (1.54)
Empty Cell	Without PWS	N = 42	86.48 (0.79)
4	All children	N = 53	70.37 (2.12)*
Empty Cell	With PWS	N = 11	67.58 (3.78)
Empty Cell	Without PWS	N = 42	73.16 (1.93

* Denotes significant differences ($P < .001$) in EQ when condition 1 is compared to conditions 2, 3, and 4 respectively.

3.3. Frequency of Falls

No participants fell on conditions 1 or 2 and only one child with PWS registered a fall on condition 3 (see Table 4). On conditions 4 and 5 there were no differences in the proportion of falling between the groups. In condition 6, more children with PWS fell than children without PWS, $\chi^2(1) = 7.468$, $p = .006$.

Table 4. Frequency (%) of children who fell at least once in each sensory condition.

Empty Cell	**Children with PWS (*n* = 14)**	**Children without PWS (*n* = 46)**	**Total (n = 60)**	**χ^2 (*p*)**
Condition 1	0	0	0	-
Condition 2	0	0	0	-
Condition 3	1 (7.1%)	0	1 (1.7%)	3.341 (.068)
Condition 4	3 (21.4%)	4 (8.7%)	7 (11.7%)	1.689 (.194)
Condition 5	8 (57.1%)	18 (39.1%)	26 (43.3%)	1.418 (.234)
Condition 6	11 (78.6%)	17 (37.0%)	28 (46.7%)	7.468 (.006)

The frequency of repeated falls within conditions 5 and 6 was also examined (see Table 5). As a result of only two children with PWS falling in multiple trials and five total children (with PWS = 1; without PWS = 4) falling once on condition 4, this condition was not examined for repeat falling. The chi-square analysis results suggested that the difference in the proportion of repeated falls between groups for condition 5 approached significance, $\chi^2(3) = 4.630$, $p = .099$. For condition 5, there were 25 falls registered with 17 falls on the first trial. In children with PWS, 17% fell on the first trial only, 33% fell on the first two trials, and 50% fell on all trials. In children without PWS, the majority fell on the first trial (63.6%), with 9.1% falling in trials 1 and 2, 18.2% on trials 1% and 3% and 9.1% on all trials. For condition 6, results of the chi-square analysis also approached significance, $\chi^2 (3) = 5.167$, $p = .076$. There were 28 falls registered with 18 falls reported on the first trial. In children with PWS, 22.2% fell on the first trial and 44% fell on all three trials. In children without PWS, 44.4% fell on the first trial but none fell on all trials.

Table 5. Repeated falls on Conditions 5 and 6 for children with and without PWS.

Empty Cell	**Condition 5**		**Condition 6**	
Empty Cell	**Children with PWS (*n* = 6)**	**Children without PWS (*n* = 11)**	**Children with PWS (*n* = 9)**	**Children without PWS (*n* = 9)**
Fell trial 1 only	1 (16.7%)	7 (63.6%)	2 (22.2%)	4 (44.4%)
Fell trial 1 & 2	2 (33.3%)	1 (9.1%)	1 (11.1%)	4 (44.4%)
Fell trial 1 & 3	0	2 (18.2%)	2 (22.2%)	1 (11.1%)
Fell all three trials	3 (50.0%)	1 (9.1%)	4 (44.4%)	0
χ^2 (*p*)	6.965 (0.073)		6.800 (0.079)	

Note: On condition 5, 1 child with PWS and 7 children without PWS fell but not on the first trial; On condition 6, 1 child with PWS and 8 children without PWS fell but not on the first trial.

4. Discussion

To our knowledge, the present study is the first to quantify and compare sensory reception and integration skills in children with and without PWS. Our results show that when compared with children with obesity but without PWS, children with PWS exhibit higher levels of sway indicating less postural control. Additionally, children with PWS were unable to adapt to

repeated exposure to sensory conditions that required the vestibular system to serve as the primary sensory input for maintaining upright balance.

Previously, adults with PWS were measured for standing balance using a static force plate under an eyes-open condition (Capodaglio et al., 2011a). Individuals with PWS demonstrated significantly higher sway in the anterior-posterior and medial-lateral directions when compared to adults with normal weight and adults with non-syndromal obesity (Capodaglio et al., 2011a). Similarly, two studies (Cimolin et al., 2011, Rubin et al., 2023) demonstrated greater displacement of the center of pressure in adults with PWS compared to those with obesity. The results of the present study are consistent with those previously shown in adults; children with PWS demonstrate higher levels of sway when compared to children with obesity as indicated by differences in the mean CES.

Hypotonia was identified as a contributing factor to poor balance in adults with PWS (Galli et al., 2011). Training programs aimed at improving use of a hip strategy to control sway and the strength of ankle agonist and antagonist muscles were postulated for potential to improve balance (Galli et al., 2011). However, previous studies showed no improvement in balance in adults with PWS when only muscle strengthening strategies were used (Capodaglio et al., 2011) suggesting balance impairments could be related to sensorimotor integration impairments.

The CES reflects overall sensory contributions to standing balance while the individual condition EQs further isolate the system(s) that are being used to maintain upright balance. For example, the sensory system primarily used to maintain upright balance in conditions 1, 2, and 3 in which the support surface is stable is the somatosensory system whereas input from the visual and vestibular systems (conditions 4, 5, and 6) is more heavily weighted when the support surface is unstable. It is therefore the system we tend to rely on most when the surface below us is stable and vision is either absent or disrupted. Research has shown that the relative weighting of incoming sensory information in healthy adults standing on a stable surface is as follows: 70% somatosensory system, 20% vestibular system, and 10% visual system (Peterka, 2002, Peterka and Loughlin, 2004). Conversely, when the surface is unstable, sensory information is re-weighted or adjusted so that the system most responsible for maintaining upright balance or responding to a postural disturbance is the vestibular system (60%). The weighting of visual system inputs also increases to 30% while somatosensory system inputs are weighted much lower at 10% (Peterka, 2002, Peterka and Loughlin, 2004). Developmental research has also shown that somatosensory function has reached maturity by as early as 5 to 8 years of age, with visual and vestibular function maturing around 12 years and 15 to 17 years, respectively (Sinno et al., 2020). Garcia-Rojas et al. (2013) also found that children between the ages of 7 and 11 years exhibit a greater dependence on the somatosensory system to control standing posture.

The results of the present study indicated that both groups demonstrated increased sway between conditions 1 and 4. Manipulation of one sensory system led to higher levels of sway as indicated by lower EQs in condition 2 (vision absent), condition 3 (vision sway-referenced), and condition 4 (somatosensory inputs manipulated) when compared to condition 1 (all sensory systems available). No appreciable difference in the EQs was evident, however, between conditions 2 (absent vision, stable support surface) and 3 (sway-referenced vision, stable support surface), suggesting that both groups of children were able to rely on somatosensory inputs to maintain standing balance when vision alone was manipulated. Conversely, manipulating somatosensory inputs produced higher levels of sway than when vision alone was manipulated (i.e., condition 4). This finding is consistent with previous results in children with DCD when standing on a compliant surface (Garcia-Rojas et al., 2013), and previous studies in

adults with PWS, in which manipulating vision was not a main determinant of balance when the surface below was stable (Cimolin et al., 2011, Peterka and Loughlin, 2004).

Differences between the two groups were also evident when the frequency of falls (greater in PWS) was compared for conditions 4 (somatosensory inputs manipulated), 5 (vision absent and somatosensory inputs manipulated), and 6 (vision sway-referenced and somatosensory inputs manipulated). In condition 4 the weighting of sensory inputs shifts to the visual and vestibular systems while in conditions 5 and 6 the weighting of sensory inputs shifts primarily to the vestibular system. The higher frequency of repeated falls in the children with PWS in conditions 5 and 6 is particularly noteworthy because it suggests that despite repeated exposure/practice, the children with PWS were unable to successfully shift the weighting of sensory input to the vestibular system in the absence of vision and manipulation of the somatosensory system (condition 5) or resolve the sensory conflict created in condition 6 as a result of manipulating visual and somatosensory input when compared to children without PWS. Oster and Zhou (2022) have also identified abnormalities in the use of vestibular inputs to control balance in pediatric patients with ASD based on their SOT® findings and the results of additional vestibular diagnostic tests (e.g., videonystagmography, rotary chair, vestibular evoked myogenic potentials). Similar results have been obtained in young adults with PWS (Rubin et al., 2023).

It is likely that the overall lack of physical activity observed in children who are overweight/obese (Elmesmari et al., 2018) and children with PWS (Butler et al., 2007, Castner et al., 2014) is detrimental to the development of static and dynamic postural control. Data show that children, including those with obesity, who engage in more physical activity of moderate intensity, demonstrate better balance (DuBose et al., 2018). Recently, engagement in a 4-week neuromuscular training program that included progressive balance activities performed on unstable surfaces and eyes closed environments resulted in significant improvements in static and dynamic postural control in children with excess body weight (Guzman-Munoz et al., 2020). Limited engagement in sensorimotor experiences among children with PWS may contribute to a vicious cycle in which a child develops poor balance and/or engages in less physical activity, which, in turn, reduces their engagement in sensorimotor experiences, further delaying the development of important postural control strategies.

Collectively, these findings suggest that movement-based interventions that systematically manipulate sensory system inputs would be beneficial for children with and without PWS. Performing different activities with the eyes closed or engaged (e.g., tracking or catching objects while standing or moving) require greater input from the somatosensory and vestibular systems (Horak, 2006, Rose, 2010). Similarly, performing activities on unstable surfaces with the eyes open, engaged, and/or closed further challenges the peripheral and central nervous system by requiring further re-weighting of sensory inputs to prioritize visual and vestibular inputs when the eyes are open and vestibular inputs when the eyes are closed or engaged.

To date, most interventions have largely focused on improving muscular strength and aerobic endurance in individuals with PWS with little attention being paid to improving the multiple dimensions of balance (Capodaglio et al., 2011). Despite the fact that the intervention designed by Rubin et al. (2019) included activities performed on unstable surfaces, as well as activities requiring visual tracking or catching of objects while standing or moving, the intervention was not specifically designed to target the multiple dimensions of balance (Rubin et al., 2019). Evaluating the efficacy of tailored interventions for dynamic and static balance in those with PWS should be pursued. As individuals with PWS present with increased risk for osteoporosis (van Nieuwpoort et al., 2018) or low bone density in children (Rubin et al., 2013), fall prevention is an important health outcome in this population.

4.1. Limitations

While these results are novel they are not without limitations. The first limitation is the small overall sample size that was further reduced in conditions 5 and 6. While trained research staff made efforts to familiarize the participants with each condition on the SOT®, these two conditions were particularly challenging and resulted in less participants completing them. One test administrator would have been ideal; however, as this is a machine standardized test, the use of four test administrators likely resulted in little variability in testing procedures between participants. While the SOT® identifies impairments in the central processing and weighting of information from three different sensory systems, translating this information into motor responses to sustain upright balance involves other factors that were not accounted for in this study. Additional neural factors include information processing speed and the generation of a timely motor response to maintain the center of mass within the base of support and prevent a loss of balance. The generation of an appropriate motor response depends upon the level of muscle strength available and muscle power. Individuals with PWS exhibit low muscle strength and power (Pamukoff et al., 2020). Specifically, in adults with PWS a lower rate of torque development was demonstrated in the upper and lower leg muscles (Pamukoff et al., 2020, Rubin et al., 2023). Early rate of torque development was defined as the torque (force) production in the first 100 ms from the onset of muscle contraction. This lower torque generation could be a contributing factor to the higher incidence of falls in children with PWS in the more challenging sensory conditions failing to generate a timely response. Additionally, anthropometric characteristics (e.g., height, weight, BMI) may influence postural control but previous research in children has yielded conflicting results thus far (McGraw et al., 2000, Peterson et al., 2006, Cumberworth et al., 2007). Last, sensory abnormalities in somatosensory (pain and tactile) perception have previously been reported in people with PWS and ASD but the extent to which they contribute to postural control and sensory integration, in particular, has yet to be determined (Brandt & Rosen, 1998; Priano et al., 2009).

5. Conclusion

Children with PWS present increased degree of sway and lower postural control compared to children with obesity but without PWS. Potentially, decreased ability to process information from the somatosensory and vestibular systems contribute to this lower postural control. The use of computerized dynamic posturography or stabilometry as a tool to evaluate and quantify postural control in children with PWS is recommended as a means to better identify the type of sensory strategies used to maintain standing balance in altered sensory environments often encountered in every day life. Additional diagnostic tests capable of identifying peripheral sensory impairments in the visual, somatosensory and vestibular systems as well as the motor system should also be included as part of the clinical management of PWS. It is further recommended that subsequent neuromuscular training programs designed for children with PWS include a specific focus on improving static and dynamic postural control in altered sensory environments, in addition to muscle strength and power.

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Declaration of Competing Interest

None.

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Data Availability

Data will be made available on request.

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