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Relationship between sensory processing and Autism Spectrum Disorder-like behaviors in Prader-Willi Syndrome

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Abstract:	The relationship between sensory processing and ASD-like and associated behaviors in patients with Prader-Willi Syndrome (PWS) remains relatively unexplored. Examining this relationship, 51 adults with PWS were administered the Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS), Short Sensory Profile (SSP-J), Food-Related Problem Questionnaire (FRPQ), and Aberrant Behavior Checklist (ABC-J). Based on SSP-J z-scores, participants were classified into three severity groups. Analysis of variance was performed to compare the behavioral scores of these three groups. Statistically significant group differences were observed in PARS (p=.006, η_p^2=.194) and ABC-J (p=.006, η_p^2=.193) scores. Our findings suggest that the level of sensory processing may predict ASD-like and aberrant behaviors in adults with PWS, implying the importance of a proper assessment for early intervention.

Title

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Abstract

The relationship between sensory processing and ASD-like and associated behaviors in patients with Prader-Willi Syndrome (PWS) remains relatively unexplored. Examining this relationship, 51 adults with PWS were administered the Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS), Short Sensory Profile (SSP-J), Food-Related Problem Questionnaire (FRPQ), and Aberrant Behavior Checklist (ABC-J). Based on SSP-J z-scores, participants were classified into three severity groups. Analysis of variance was performed to compare the behavioral scores of these three groups. Statistically significant group differences were observed in PARS (p=.006, η_p^2=.194) and ABC-J (p=.006, η_p^2=.193) scores. Our findings suggest that the level of sensory processing may predict ASD-like and aberrant behaviors in adults with PWS, implying the importance of a proper assessment for early intervention.

Keywords: Prader-Willi Syndrome; Sensory processing; ASD-like behaviors; Aberrant behaviors.

Introduction

Prader-Willi Syndrome (PWS) is a contiguous genetic syndrome caused by either a paternal deletion (DEL) of 15q11-q13, observed in 70% of patients, or maternal uniparental disomy 15 (mUPD; when both copies of chromosome 15 are maternally inherited), observed in 25% of patients (Bolton et al., 2001; Chamberlain & Brannan, 2001; Dimitropoulos & Schultz, 2007; Dykens, 2004; Veltman et al., 2004). The main clinical symptoms of PWS are neonatal hypotonia, intellectual disability, hyperphagia, progressive obesity, and hypogonadism (Bailey et al., 2002; Cassidy & Driscoll, 2009). In addition to hyperphagia, individuals with PWS exhibit several behavioral and psychiatric symptoms, including Autism Spectrum Disorder (ASD)-like behaviors (Arron, Oliver, Moss, Berg, & Burbidge, 2011; Klabunde et al., 2015). Several studies have been conducted to identify underlying genotypic differences between individuals with mUPD and DEL subtypes. Individuals with PWS of the mUPD subtype have exhibited a greater prevalence of ASD-like behaviors, such as compulsive, ritualistic, and repetitive behaviors, than did those of the DEL subtype (Sinnema et al., 2011; Soni et al., 2007; Wigren & Hansen, 2005). These findings with reference to the susceptibility of ASD-like behaviors in individuals with mUPD imply that maternally active gene(s) may lie in chromosome 15q11-q13 (Dykens, Maxwell, Pantino, Kossler, & Roof, 2007; Ogata et al., 2014; Vogels, Matthijs, Legius, Devriendt, & Fryns, 2003; Wigren & Hansen, 2005). This possibility is consistent with the fact that the most common cytogenetic abnormality in individuals with ASD, detected in 1–3%, is the maternally inherited 15q11-q13 duplication (Baker, Piven, Schwartz, & Patil, 1994).

There has been considerable debate regarding the basic behavioral characteristics of individuals with ASD. The core symptoms of ASD include difficulties with social interaction and communication, as well as restricted and repetitive behaviors. Beyond these, sensory processing impairment may also be one of the core deficits underlying ASD. The Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5) lists “hyper- or hyporeactivity to sensory input or unusual interest in sensory aspects of the environment” as one of the four restricted, repetitive patterns of behavior. The other three are stereotyped or repetitive motor movements, insistence on sameness, and restricted and fixated interests. Ample evidence suggests that 45–90% of individuals with ASD show high rates of sensory processing impairments (Ben-Sasson et al., 2009) exceeding one standard deviation (Dimitropoulos, Feurer, Butler, & Thompson, 2001; Dykens, Cassidy, & King, 1999; Dykens, Sutcliffe, & Levitt, 2004; Dykens & Roof, 2008; Jauregi, Laurier, Copet, Tauber, & Thuilleaux, 2013; Symons, Butler, Sanders, Feurer, & Thompson, 1999). Atypical responses characteristic of ASD have been observed even in high-functioning individuals (Einfeld et al., 2006), implying that poorer sensory processing is not always associated with a lower IQ. Sensory processing impairments, such as over-responsivity to tactile and auditory non-target inputs, constitute prodromal signs that parents can use to detect the presence of developmental disorders in their children for the first time. The early emergence of sensory processing impairments in toddlers often indicates that such disorders will influence a child’s adaptive behaviors from an early stage of development (Ben-Sasson, Carter, & Briggs-Gowan, 2009). The comorbidity of PWS with ASD-like behaviors implies that sensory processing impairments are rooted in several autistic and allied behavioral symptoms.

In addition to ASD-like behaviors, PWS-specific food-related problems in relation to sensory processing impairments are worth exploring. These problems seem to be different from the picky-eating phenomenon seen in ASD. This is because hyperphagia in PWS is evidently linked to a constant insatiable appetite, perhaps due to dysfunction of the satiety control system (Lindgren et al., 2000). Therefore, compared with picky eating in ASD, food-related problems in individuals with PWS are severe and diverse, including food stealing, lying, and pica (Hiraiwa, Maegaki, Oka, & Ohno, 2007). Taking such uniqueness of food-related behaviors in individuals with PWS into account, a thorough analysis with regard to the association between these behaviors and sensory processing impairments should be conducted.

To date, the relationship between sensory processing impairments and ASD-like behaviors in PWS has yet to be elucidated. One of a few studies that investigated sensory processing impairments in individuals with PWS was conducted by Takahashi, Ihara, and Ogata (2019). They reported that approximately 75% of patients with PWS demonstrated impairments in sensory responsiveness. As far as general, non-ASD-like, psychiatric symptoms (depressed mood, general anxiety, mania/hyperactivity, obsessive compulsive behavior, social avoidance) are concerned, Royston et al. (2020) found that auditory sensory processing impairments were associated with psychiatric symptoms in individuals with Williams Syndrome, but not in individuals with PWS. However, the relationship between sensory processing impairments and ASD-like and associated behaviors remains largely unexamined. As for PWS, the relationship has never been examined.

When investigating the association between maladaptive behaviors and sensory processing impairments, two factors should be considered: developmental trajectory and genotypic differences. It has been argued that problem behaviors, such as temper tantrums, compulsions, self-injurious behaviors, and ASD-like behaviors, follow a non-linear trajectory (Dimitropoulos et al., 2001; Dykens et al., 2004; Jauregi et al., 2013). For example, Ishii et al. (2017) reported that ASD-like behaviors follow a marked trend of aggravation beginning at approximately 18 years of age. Considering the transition of ASD-like behaviors with development, research should focus on adults as well as children and adolescents. Studies have reported a higher risk of ASD-like behaviors in individuals with the mUPD subtype than with the DEL subtype (Sinnema et al., 2011; Soni et al., 2007; Wigren & Hansen, 2005).

With regard to genotypic differences in sensory processing, Takahashi, Ihara, and Ogata (2019) reported a marginal difference in auditory filtering, in which individuals with mUPD showed a trend towards impairment compared with individuals with DEL. As such, the influence of genotype on the relationship between sensory processing impairments and ASD-like behaviors merits investigation.

To the best of our knowledge, this study is the first attempt to address the relationship between sensory processing, on the one hand, and ASD-like, aberrant, and compulsive eating behaviors, on the other, in adults with PWS. A high incidence of sensory processing impairments has already been indicated in individuals with ASD, whose common cytogenetic abnormality is duplication of the 15q11-q13 PWS/AS region. The hypothesis is that sensory processing impairments are cardinal deficits leading to a variety of maladaptive behaviors in individuals with PWS, as indicated in ASD. First, this study attempted to characterize sensory processing impairments in adults with PWS. Second, the study aimed to investigate the association between sensory processing impairments and other behavioral symptoms, including aberrant and food-related behaviors, as well ASD-like behaviors such as interpersonal skills, communication, and obsession. Finally, the study aimed to examine the differences between individuals with DEL and individuals with mUPD with respect to sensory processing and other maladaptive behaviors.

Method

Ethical Approval, Participants, and Procedure

This study was commenced after being assessed and approved by the Institutional Review Board of our university. All research procedures conformed to the World Medical Association Declaration of Helsinki (adopted in October 2013 in Brazil). Before the data were collected, participants or their parents provided informed consent for behavioral and psychiatric assessments, and cytogenetic and/or molecular-genetic studies. In total, 51 Japanese participants with PWS (aged 17 to 48 years) participated in this study. Diagnoses had already been made for all patients based on fluorescence *in situ* hybridization or the methylation test prior to this study. The participants comprised 31 male and 20 female individuals, including 41 patients with DEL and 10 patients with mUPD. The assessor who collected the data was blinded to the genetic status of each patient. Before administering a comprehensive set of behavioral measures, the IQ of each participant was measured using the Japanese version of the Wechsler Intelligence Scale (Wechsler, 1991, 1997; Japanese WISC-III Publication Committee, 1998; Japanese WAIS-III Publication Committee, 2006). The assessor collected behavioral data over three to six sessions for each participant. Most behavioral measures applied in this study were originally designed to be self-administered or informant-based. Due to participants' difficulty with instructions and low level of cognitive ability, the assessor met with participants and their parents in person to answer any questions while the behavioral assessments were completed by the participants. It was thus expected that the quality of data obtained in this study would be superior to that obtained using mail questionnaires.

Measures

Sensory Processing

The sensory processing ability of all participants was measured using the Japanese version (SSP-J) (Tsujii et al., 2015) of the Short Sensory Profile (SSP) (Dunn, 1999). The SSP-J is a 38-item caregiver questionnaire constructed to examine the frequency of sensory-processing behaviors in a child. Raw scores were allocated using a five-point Likert scale (*always, five; frequently, four; occasionally, three; seldom, two; or never, one*) (Tsujii et al., 2015).

Attention should be paid to the following difference between SSP and SSP-J: lower scores represent worse sensory processing in SSP, whereas higher scores represent worse sensory processing in SSP-J. The questionnaire comprised seven subscores: Tactile Sensitivity, Taste/Smell Sensitivity, Movement Sensitivity, Underresponsive/Seeks Sensation, Auditory Filtering, Low Energy/Weak, and Visual/Auditory Sensitivity. A higher total score indicated more severe impairment. The internal reliability of each subsection including the seven subscores and total SSP-J score in 1,441 typically developing children in Japan was between 0.69 and 0.84. Moreover, no significant difference in Cronbach's coefficient alpha was found between subjects aged 3–10 or those aged 11–82 (Tsujii et al., 2015). Hence, SSP-J is applicable to the adult population.

According to the criteria proposed by McIntosh et al. (1999), the raw scores of eight subsections were converted to standardized z-scores. In the child's responses to sensory experiences, "Typical Performance" indicated z-scores above -1.00, "Probable Difference" indicated those from -1.00 to -2.00, and "Definite Difference" indicated those below -2.00. This classification system, made up of three categories (Typical Performance, Probable Difference, and Definite Difference), has been used in previous studies (Caron, Schaaf, Benevides, & Gal, 2012; Nadon, Feldman, Dunn, & Gisel, 2011).

Behavioral Assessment

ASD-like Symptomatology. To assess ASD-like symptomatology, the Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS) (Adachi et al., 2006; Kamio et al., 2006) was administered. This rating scale was developed as a questionnaire to measure the degree of autistic and allied behaviors in Pervasive Developmental Disorders (PDDs). When assessing the severity of current ASD-like behaviors in this study, a 33-item version for adolescents and adults was applied. The PARS for this population was divided into five clinical subscores including Interpersonal Skills (six items), Communication (seven items), Obsession (six items), Problematic Behaviors (11 items), and Hypersensitivity (three items). Reliability and validity of the PARS were established for both the childhood items (Adachi et al., 2006) and the adolescent and adult items (Kamio et al., 2006).

Aberrant Behaviors. The extent of participants' maladaptive and problematic behaviors was measured based on the Aberrant Behavior Checklist Japanese Version (ABC-J) (Aman, Singh, & Ono, 2006). The ABC-J included 58 items, which took 10–15 minutes to complete. All items consisted of five categories: a) Irritability and Agitation, b) Lethargy and Social Withdrawal, c) Stereotypic Behavior, d) Hyperactivity and Noncompliance, and e) Inappropriate Speech. The ABC is confirmed to be an effective tool in evaluating the severity of behavioral manifestations in individuals with intellectual disability (Shedlack, Hennen, Magee, & Cheron, 2005) and ASD (Brinkley et al., 2007). This tool was also applied to measure the effects of treatment (Schroeder, Rojahn, & Reese, 1997; Shedlack et al., 2005). The reliability and validity of the Japanese version of the ABC were established by Aman et al. (2006). The ABC has been used for the purpose of evaluating the severity of problem behaviors (Clarke, Boer, Chung, Sturmey, & Webb, 1996) and the effect of pharmacotherapy (Shapira, Lessig, Lewis, Goodman, & Driscoll, 2004) in individuals with PWS.

Food-related Behaviors. To assess food-related behaviors, the Food-Related Problem Questionnaire (FRPQ) was used. This is an informant-based questionnaire formed uniquely for evaluating the severity of eating behaviors in individuals with PWS. The FRPQ comprises 16 items, including three subscales: preoccupation with food (P), impairment of satiety (S), and other food-related negative behaviors (N). The FRPQ has sufficiently robust psychometric properties, in terms of test-retest and inter-rater reliability, concurrent and criterion validity, and internal consistency (Russel & Oliver, 2003).

Statistical analyses

IBM SPSS Statistics for Windows, version 20.0 (IBM Corp., Armonk, NY, USA) was used to analyze the data. According to the z-scores of the SSP-J raw scores, participants were classified into three performance categories: Typical Performance (z-score above -1.00), Probable Difference (z-score from -1.00 to -2.00), and Definite Difference (z-score below -2.00). To compare the level of sensory processing with the severity of other behavioral symptoms, one-way analysis of variance (ANOVA) tests were conducted to assess differences in scores of PARS, ABC-J, and FRPQ among the three performance groups. To assess the differences between individuals with DEL and individuals with mUPD, two-tailed *t*-tests were conducted on raw scores of the eight SSP-J subsections. A *p*-value of 0.05 or less was regarded as statistically significant for all statistical tests.

Results

Descriptive Statistics: Sensory Processing Differences

Performance classifications based on the z-scores of SSP-J raw scores are presented in Table 1. SSP-J test results were as follows: 23.5% (n = 12) of the adults with PWS obtained Typical Performance scores (z-score above -1.00), 41.2% (n = 21) obtained Probable Difference scores (z-score from -1.00 to -2.00), and 35.3% (n = 18) obtained Definite Difference scores (z-score below -2.00). More detailed examination of the seven subscores revealed that the Low Energy/Weak subsection yielded the highest reported Definite Differences (z-score below -2.00) (43.1%, n = 22). Other SSP-J subsections that bore higher percentages of Definite Differences (z-score below -2.00) included Movement Sensitivity (27.5%, n = 14) and Underresponsive Sensitivity (15.7%, n = 8). Conversely, lower percentages of Definite Difference (z-score below -2.00) were observed in the following subsections: Taste/Smell

Sensitivity (0%, n = 0), Tactile Sensitivity (7.8%, n = 4), Auditory Filtering (7.8%, n = 4), and Visual/Auditory Sensitivity (9.8%, n = 5).

Table 2 shows the patient characteristics of the three groups based on the results of total SSP-J scores. A one-way ANOVA did not reveal any statistically significant differences among the three groups with regard to age, body mass index, or IQ.

Sensory Processing and ASD-Like Behaviors

To compare the level of sensory processing with the severity of ASD-like behaviors in adults with PWS, one-way ANOVAs were conducted to assess differences in PARS scores among the three groups which were categorized based on the SSP-J results: Typical Performance (z-score above -1.00), Probable Difference (z-score from -1.00 to -2.00), and Definite Difference (z-score below -2.00) (see Table 3). Statistically significant differences were detected in the PARS Total Score (p=.006) and Communication subscore (p<.001). Post-hoc Tukey's tests demonstrated that adults with PWS with Definite Difference (z-score below -2.00) scored higher than those with Typical Performance (z-score above -1.00) with regard to PARS Total Score (p=.004) and Communication subscore (p<.001) (see Figure 1). There were no statistically significant differences among the three groups with respect to Interpersonal Skills, Obsession, Problematic Behaviors, and Hypersensitivity. These analyses revealed that greater ASD-like behaviors were found in individuals with more a severe level of sensory processing impairment.

Sensory Processing and Aberrant Behaviors

The relationships between sensory processing and aberrant behaviors were examined using one-way ANOVAs to assess differences in ABC-J scores among the three performance groups (see Table 3). Statistically significant differences were observed in the ABC-J Total

Score (*p*=.006) and four subscores (Irritability and Agitation, *p*=.003; Lethargy and Social Withdrawal, *p*=.005; Stereotypic Behavior, *p*=.030; Inappropriate Speech, *p*=.003) in the ABC-J. In all ABC-J scores, individuals with Definite Difference (z-score below -2.00) in sensory processing exhibited the most severely aberrant behaviors. Those with Probable Difference (z-score from -1.00 to -2.00) were moderately aberrant, and those with Typical Performance (z-score above -1.00) exhibited the lowest level of aberrations. Post-hoc Tukey's tests revealed statistically significant differences in aberrant behaviors as follows: members of the Definite Difference (z-score below -2.00) group scored higher than those of the Typical Performance (z-score above -1.00) group in the ABC-J Total Score (*p*=.009) and four of the five subscores (Irritability and Agitation, *p*=.005; Lethargy and Social Withdrawal, *p*=.035; Stereotypic Behavior, *p*=.040; Inappropriate Speech, *p*=.003). Members of the Definite Difference (z-score below -2.00) group scored higher than those of the Probable Difference (z-score from -1.00 to -2.00) group in the ABC-J Total score (*p*=.027), Irritability and Agitation subscore (*p*=.026), and Lethargy and Social Withdrawal subscore (*p*=.007) (see Figure 1). To sum up, individuals with the most severe sensory processing impairments also had more severe problematic behavior.

Sensory Processing and Food-related Behaviors

In order to examine whether there were differences in PWS-specific food-related behaviors based on the level of sensory processing impairment, one-way ANOVAs were conducted to investigate differences in the FRPQ scores among the three performance groups. No statistically significant differences were noted in Total Scores or the three subscores of the FRPQ (Preoccupation with Food (P), Impairment of Satiety (S), and other Food-related Negative Behaviors (N)) (see Table 3). Therefore, PWS-specific food-related behaviors did not differ based on the level of sensory processing.

Genotypic Differences

Multiple *t*-tests were used to assess differences in raw scores of the eight SSP-J subsections between individuals with DEL and individuals with mUPD. As demonstrated in Table 4, statistically significant differences were observed in the Auditory Filtering subsection, in which individuals with mUPD demonstrated a significantly higher score (p=.041), but this was not the case for other subsections or the Total Scores.

T-tests were conducted to assess genotypic differences in PARS, ABC-J, and FRPQ (see Table 4). Adults with mUPD scored higher with regard to the PARS Total Score (p=.002), three PARS subscores (p=.013 in Interpersonal Skills, p=.048 in Communication, and p=.004 in Problematic Behaviors), ABC-J Total Score (p=.002), and all five ABC-J subscores (p=.011 in Irritability and Agitation, p<.001 in Lethargy and Social Withdrawal, p=.001 in Stereotypic Behavior, p=.001 in Hyperactivity and Noncompliance, and p=.046 in Inappropriate Speech). Members of the mUPD adult group scored lower in the FRPQ Total Score (p=.030) and two FRPQ subscores (p=.049 in FRPQ-P and p=.018 in FRPQ-N). Medians and *p*-values are presented in Table 4.

Discussion

First, this study examined whether sensory processing was impaired in a sample of adults with PWS. In this study, more than 75% of adults with PWS exhibited impairments in sensory processing ability, while 23.5% of the sample qualified with Typical Performance (z-score above -1.00) on the basis of the SSP-J Total Score. Individual examination of the seven subsections of the SSP-J revealed that the most profound impairment was in the Low Energy/Weak subsection. Thus, the most impaired domain of sensory processing was the ability to contract muscles, maintain sufficient muscle tone, and control proper posture. Likewise, severe impairment was observed in the ability to respond to touch stimuli (Tactile Sensitivity) and movement experiences (Movement Sensitivity), and to modulate the level of awareness of sensory events (Underresponsive/Seeks Sensation). In contrast, less severe impairments were noted in responses to taste, smell (Taste/Smell Sensitivity), sights, and sounds (Visual/Auditory Sensitivity).

Our data suggest that adults with PWS experience sensory processing impairments. To examine whether these impairments were related to behavioral problems, such as ASD-like, food-related, and aberrant behaviors, three performance groups of sensory processing were compared with regard to PARS, FRPQ, and ABC-J scores.

Greater severity of ASD-like behaviors was found in individuals with a greater severity of sensory processing impairments. Among the five PARS subscores, the most striking feature associated with ASD-like behaviors in adults with PWS was in the Communication subscore. Compared with adults with PWS with Typical Performance (z-score above -1.00), those with Definite Difference (z-score below -2.00) and those with Probable Difference (z-score from -1.00 to -2.00) in the SSP-J categories were profoundly impaired in communication; thus, ASD-like communication problems in adults with PWS may be reflective of sensory processing impairments. Nevertheless, results from inter-group comparisons cannot prove causality between sensory processing impairments and communication. Questions remain unanswered as to whether ASD-like behaviors in general and communication problems in particular are based on sensory processing impairments or merely their concomitant phenomena. Even in ASD, a debate is still ongoing as to whether sensory processing impairments are an essential attribute or an accidental property (Ben-Sasson et al., 2009). In PWS, several factors including non-social contingencies (Didden, Korzilius, & Curfs, 2007) may underscore PWS-related ASD-like communication difficulties, such as avoiding unpleasant stimuli, reacting to and/or resisting unpleasant sensory experiences, and induction by irrelevant sensory information. A systematic aggregation of evidence is needed to clarify whether sensory symptoms (i.e., temper tantrums, compulsiveness, ritualistic behaviors, skin-picking behaviors, and autistic-like behaviors) should be regarded as core behavioral features of PWS.

Our data also suggest a relationship between sensory processing and aberrant behaviors. Greater severity in sensory processing impairment was associated with greater severity in aberrant behaviors. To date, the relationship between problematic behaviors and impaired sensory responses has mainly been investigated in younger groups, such as children with ASD (Hilton et al., 2010; O'Donnell, Deitz, Kartin, Nalty, & Dawson, 2012; Tomchek & Dunn, 2007) and those with Williams Syndrome (Glod, Riby, & Rodgers, 2020; Royston et al., 2020). By building on these studies, this research highlights new data concerning an adult population with the rare genetic syndrome PWS.

The relationship between sensory processing and maladaptive behaviors, specifically in the contrast between food-related problems and non-food-related problems, is important. Aside from PWS, Zickgraf et al. (2020) reported that rigidity and oral texture sensitivity were statistically significantly correlated with selective eating in both ASD and non-ASD samples. Engel-Yeger et al. (2016) also found significant correlations between sensory processing impairments and eating problems in individuals with intellectual disability. However, in this study of individuals with PWS with ASD-like behaviors and intellectual disability, there were no statistically significant relationships between sensory processing and PWS-specific food-related behaviors, such as preoccupation with food, impairment of satiety, and miscellaneous food-related problems. In contrast, a more severe level of non-food-related behaviors, such as ASD-like and aberrant behaviors, was observed in individuals with more severe sensory processing impairments. The trajectory of PWS-specific food-related behaviors has already been highlighted. According to Ishii et al. (2017), food-related behaviors do not typically change after 18 years of age, whereas ASD-like and aberrant behaviors worsen following this transitory stage. These findings support the opinion of Pignatti et al. (2013) concerning the results of statistical clustering. They proposed that hyperphagia and allied maladaptive eating behaviors belong to a statistical cluster distinct from other clusters that include compulsive symptoms and destructive behaviors. In demonstrating greater maladaptive behaviors in individuals with the most severe level of sensory processing impairment, this study strengthens the perspective that the problem behaviors of PWS include two different groups: food-related problems and non-food-related problems.

With regard to an intergenotypic comparison of sensory processing, adults with PWS with mUPD were more severely impaired than were those with DEL in their ability to select and screen out sounds (Auditory Filtering). Intergenotypic differences were also noted in ASD-like and aberrant behaviors; adults with PWS with mUPD were more severely impaired than were those with DEL **(**Ishi et al., 2017; Ogata et al., 2014). The possibility that overwhelming of the auditory senses due to impaired filtering ability may cause maladaptive behaviors has been suggested in various neurodevelopmental disorders, including ASD (Baranek et al., 2002; Ben-Sasson et al., 2007; Lane, Young, Baker, & Angley, 2010; Lane, Reynolds, & Dumenci, 2012) and Down syndrome (Will et al., 2019), but not in PWS. Further research is needed to address whether impaired auditory filtering may lead to maladaptive behaviors in individuals with PWS, and whether this is more profound in those with the mUPD subtype. Moreover, the factors underlying the reversal of the FRPQ results of adults with mUPD and DEL with regard to food-related problem behaviors (i.e., such behaviors were less prevalent in adults with mUPD than in their DEL counterparts) remain unresolved. Although the aforementioned uniqueness of food-related problem behaviors has been considered, severe impairments in auditory filtering in adults with PWS with mUPD merit further study since the severity of auditory filtering impairments were in marked contrast with less severe impairments in food-related problem behaviors.

The results of this study highlight the significance of proper evaluation of sensory processing in adults with PWS, as the majority of adults with PWS in this study exhibited sensory processing impairments. Additionally, groups with higher rates of impairment were found to have increased ASD-like and aberrant behaviors. Such sensory processing impairments can be detected in daily life settings, such as the SSP, but not via laboratory-based neurophysiological examinations. This was suggested by Priano et al. (2009), who found that electroneurographic examination, sympathetic skin response, and somatosensory evoked potentials were all within normal ranges in adults with PWS. Therefore, there is an urgent need for a comprehensive assessment focusing on sensory processing in the real-world context by means of standardized scales like the SSP-J. This is particularly true for adults with PWS with mUPD. Ample evidence has demonstrated that individuals with the mUPD subtype are at higher risk of having ASD-like social impairments (Ogata et al., 2014). Further research is needed to investigate the possibility that ASD-like social impairment in individuals with mUPD is reflective of a lower degree of auditory filtering.

From a practical point of view, this study implies the importance of early detection of sensory processing for early intervention in individuals with PWS. Alkhamra and Abu-Dahab. (2020) have suggested that early detection and intervention in terms of sensory processing impairment may assist in reducing the risk of neurobehavioral problems, including social-emotional problems, in children with hearing impairments. Equally helpful may be the early assessment of sensory processing in individuals with PWS. Indeed, caregivers of individuals with PWS tend to be concerned about conspicuous behavioral problems like temper tantrums, compulsion, and autism-like behaviors. However, such behaviors could be predicted in advance if the level of sensory processing were thoroughly examined. Therefore, an early assessment followed by a proper intervention plan in terms of sensory processing would reduce the risk of autism-like and aberrant behaviors and enhance overall functioning of individuals with PWS.

The current study has several methodological limitations. First, as this study focused on a rare genetic disorder, the sample size was small. In addition, a large difference in the number of participants existed between the two genotype groups: 41 patients with DEL and 10 patients with mUPD. Moreover, the sample consisted of patients with a large age range, between 17 and 48 years of age. Therefore, the power is limited, inevitably resulting in an inflation of type 1 error rates. Second, this study was cross-sectional rather than longitudinal. Hence, behavioral variables were not studied over time. To examine the potential causal relationship between sensory processing and other behaviors in more detail, longitudinal studies are needed to track the same cohort for a certain period. Third, the extent of comorbidities and medication use in individuals with PWS should be considered when examining the influence of sensory processing impairment on the level of ASD-like and aberrant behaviors, as these can have effects on sensory processing. Finally, the fact that the most profound impairment was in the Low Energy/Weak subsection warrants further investigation. Indeed, neonatal hypotonia is one of the main clinical features of PWS. Although the study sample was adults, rather than children, with PWS, hypotonia may have affected the severity of the Low Energy/Weak subsection. From the above, any conclusions regarding the relationship of sensory processing with ASD-like and other behaviors should be treated with caution. Future research with larger samples and collection of more detailed patient background is needed to investigate the relevance of sensory processing and behavioral disorders in individuals with PWS.

Our findings suggest that the level of sensory processing may predict ASD-like and aberrant behaviors in adults with PWS. Auditory filtering of adults with PWS with mUPD was more severely impaired than that of adults with PWS with DEL. The results of this study highlight the significance of early assessment followed by a proper intervention plan in terms of sensory processing in adults with PWS.

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Figure 1. Effects of the sensory profile of Prader-Willi Syndrome on the total and communication scores of PARS, and the total, irritability and agitation, lethargy and social withdrawal, stereotypic behavior, and inappropriate speech scores of the ABC - J. Horizontal lines above the bars indicate significant differences between groups (*p < .05; **p < .01).

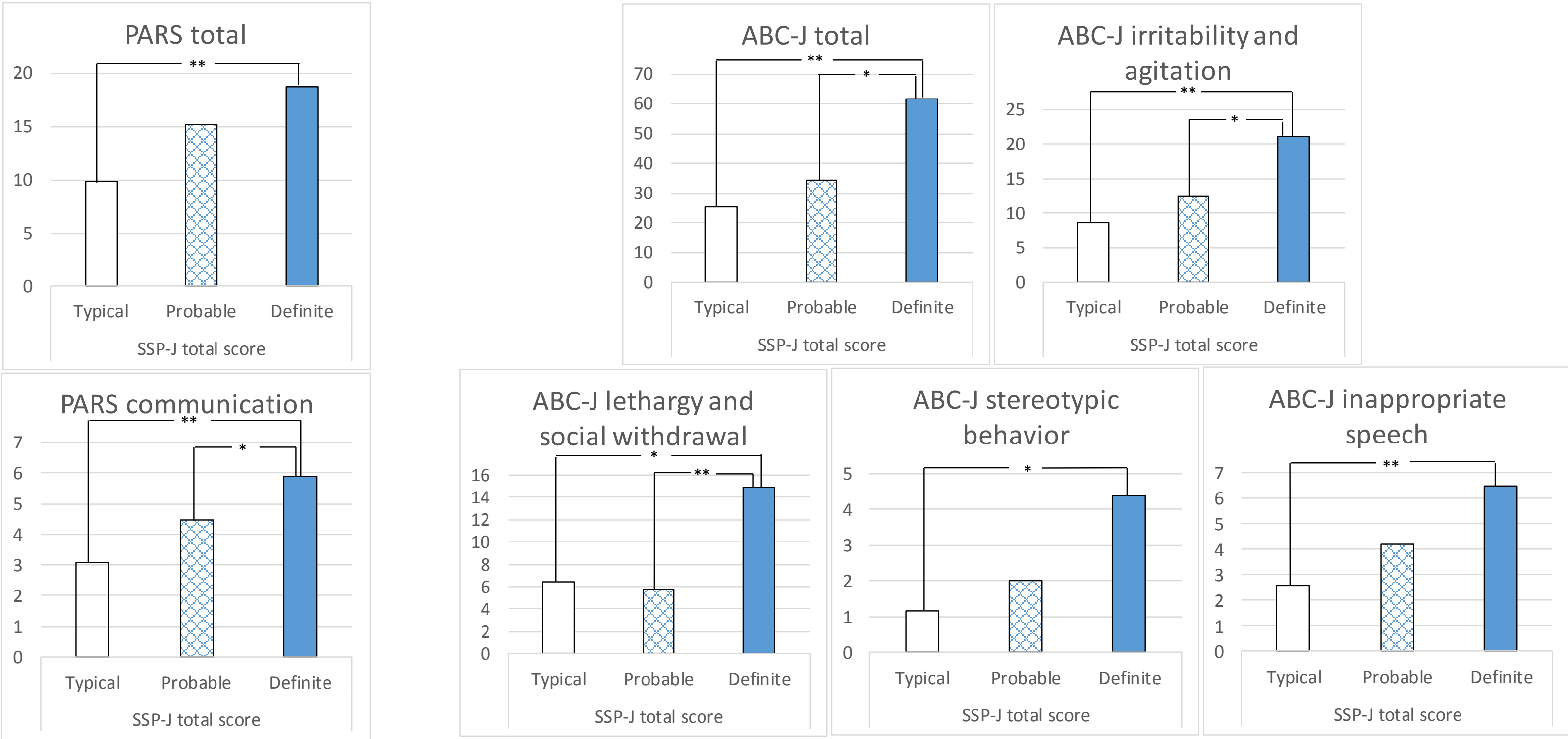

Table 1

Performance Classification of the SSP-J Subsections in total 51 participants with PWS

	SSP-J Categories		
	Typical N(%)	Probable N(%)	Definite N(%)
SSP-J total	12(23.5%)	21(41.2%)	18(35.3%)
SSP-J tactile sensitivity	17(33.3%)	30(58.8%)	4(7.8%)
SSP-J taste/smell sensitivity	37(72.5%)	14(27.5%)	0(0%)
SSP-J movement sensitivity	19(37.3%)	18(35.3%)	14(27.5%)
SSP-J underresponsive sensitivity	17(33.3%)	26(51.0%)	8(15.7%)
SSP-J auditory filtering	31(60.8%)	16(31.4%)	4(7.8%)
SSP-J low energy/weak	7(13.7%)	22(43.1%)	22(43.1%)
SSP-J visual/auditory sensitivity	38(74.5%)	8(15.7%)	5(9.8%)

Note. Typical, Probable, and Definite are the three categories participants were assigned to based on the standardized z-scores of the total SSP-J score, corresponding to z-scores above -1.00, between -1.00 and -2.00, and below -2.00, respectively. SSP-J = Short Sensory Profile, Japanese version.

Table 2

Patient Characteristics in the Three Performance Groups

	Total	SSP-J categories		
		Typical	Probable	Definite
Number	51	12(23.5%)	21(41.2%)	18(35.3%)
DEL/mUPD	41/10	10/2	19/2	12/6
Male/Female	31/20	6/6	14/7	11/7
Mean age	24.98	23.17	26.19	24.78
Age range	17-48	17-31	18-46	17-48
Mean BMI	32.54	29.57	33.69	33.19
BMI range	16.10-72.23	16.10-47.46	17.29-72.23	19.17-58.12
IQ mean(N)	48.45(42)	46.80(10)	47.06(18)	51.43(14)
IQ range	39-76	39-53	39-62	39-76

Note. Typical, Probable, and Definite are the three categories participants were assigned to based on the standardized z-scores of the total SSP-J score, corresponding to z-scores above -1.00, between -1.00 and -2.00, and below -2.00, respectively. SSP-J = Short Sensory Profile, Japanese version; DEL =

paternal deletion; mUPD = maternal uniparental disomy; BMI = body mass index.

Table 3

BMI, IQ, PARS, ABC-J, and FRPQ Scores and the Results of One-Way ANOVA Using the SSP-J Categories

	Total	SSP-J categories			ANOVA interaction	
					F	*P*
		Typical	Probable	Definite		
BMI	32.54 ± 12.141	29.57 ± 9.123	33.69 ± 14.710	33.19 ± 10.808	0.469	0.628
FIQ	48.45 ± 7.967	46.80 ± 4.517	47.06 ± 5.578	51.43 ± 11.447	1.505	0.235
VIQ	55.76 ± 6.760	54.90 ± 2.923	54.22 ± 5.320	58.36 ± 9.492	1.628	0.209
PIQ	49.26 ± 8.302	47.20 ± 5.514	48.17 ± 5.044	52.14 ± 12.215	1.329	0.276
PARS total	15.20 ± 7.699	9.83 ± 5.875	15.19 ± 6.439	18.78 ± 8.328	5.789	0.006**
PARS interpersonal skills	2.65 ± 2.528	1.42 ± 0.996	2.62 ± 2.711	3.50 ± 2.771	2.603	0.084
PARS communication	4.65 ± 2.018	3.08 ± 1.505	4.48 ± 2.015	5.89 ± 1.530	9.492	0.000**
PARS obsession	2.61 ± 1.733	1.75 ± 1.485	3.05 ± 1.658	2.67 ± 1.847	2.266	0.115
PARS problematic behaviors	4.31 ± 3.513	3.00 ± 3.542	4.00 ± 2.588	5.56 ± 4.162	2.141	0.129
PARS hypersensitivity	0.98 ± 0.787	0.58 ± 0.793	1.05 ± 0.740	1.17 ± 0.786	2.209	0.121
ABC-J total	41.86 ± 34.299	25.33 ± 29.809	34.48 ± 33.271	61.50 ± 30.237	5.748	0.006**

ABC-J irritability and agitation	14.59 ± 10.980	8.67 ± 7.644	12.43 ± 11.733	21.06 ± 8.947	6.416	0.003**
ABC-J lethargy and social withdrawal	9.16 ± 9.739	6.42 ± 13.007	5.76 ± 6.147	14.94 ± 8.370	5.894	0.005**
ABC-J stereotypic behavior	2.65 ± 3.632	1.17 ± 2.725	2.00 ± 2.864	4.39 ± 4.368	3.777	0.030*
ABC-J hyperactivity and	10.82 ± 10.514	6.42 ± 7.179	10.10 ± 12.173	14.61 ± 9.375	2.400	0.102
ABC-J inappropriate speech	4.63 ± 3.340	2.58 ± 2.193	4.19 ± 3.156	6.50 ± 3.330	6.387	0.003**
FRPQ total	39.35 ± 14.802	44.58 ± 9.337	41.33 ± 12.978	33.56 ± 18.170	2.452	0.097
FRPQ-P	9.67 ± 4.462	10.50 ± 5.351	10.33 ± 4.078	8.33 ± 4.187	1.260	0.293
FRPQ-S	17.39 ± 5.437	20.17 ± 2.657	17.62 ± 4.177	15.28 ± 7.185	3.201	0.050
FRPQ-N	12.22 ± 6.813	13.92 ± 5.418	13.19 ± 6.194	9.94 ± 7.981	1.629	0.207

Note. Typical, Probable, and Definite are the three categories participants were assigned to based on the standardized z-scores of the total SSP-J score, corresponding to z-scores above -1.00, between -1.00 and -2.00, and below -2.00, respectively. SSP-J = Short Sensory Profile, Japanese version; BMI = body mass index; FIQ = Full Scale Intelligence Quotient; VIQ = Verbal Intelligence Quotient; PIQ = Performance Intelligence Quotient; PARS = Pervasive Developmental Disorders Autism Society Japan Rating Scale; ABC-J = Aberrant Behavior Checklist, Japanese version; FRPQ = Food-Related Problem Questionnaire; FRPQ-P = Food-Related Problem Questionnaire – preoccupation with food; FRPQ-S = Food-Related Problem Questionnaire – impairment of satiety; FRPQ-N = Food-Related Problem Questionnaire – other food-related negative behaviors.

*$p < .05$. **$p < .01$

22 Table 4

23 *SSP-J, PARS, ABC-J, and FRPQ Scores in the Groups and Comparison of the Two Genotypes*

	Genotype				*P*-value
	DEL, N=41		mUPD, N=10		
	Median	Q1;Q3	Median	Q1;Q3	
SSP-J total	64	53.5;80.5	83	55.75;94.75	0.138
SSP-J tactile sensitivity	11	8.5;13.5	15	8.5;20.5	0.125
SSP-J taste/smell sensitivity	4	4;4.5	4	4;6	0.257
SSP-J movement sensitivity	6	3;9	7	3.75;9.25	0.657
SSP-J underresponsive	10	7;14	12	8.75;17.25	0.148
SSP-J auditory filtering	9	7;12	12	9.75;15	0.041*
SSP-J low energy/weak	17	12;23	19	15.75;24.75	0.468
SSP-J visual/auditory	6	5;7	7	6;10.75	0.071
PARS	12	9;17.5	21	18.5;26.5	0.002**
PARS interpersonal skills	1	1;3	4	2;7.25	0.013*
PARS communication	4	3;5	6	4;8	0.048*

PARS obsession	2	1;3.5	3.5	2;4.25	0.108
PARS problematic behaviors	2	1.5;5	7.5	5.75;8.75	0.004**
PARS hypersensitivity	1	0.5;1	1	0;2	0.856
ABC-J total	25	10.5;48.5	82.5	42.75;113	0.002**
ABC-J irritability and agitation	11	5;17	25.5	14.75;35.25	0.011*
ABC-J lethargy and social	4	1;10	19.5	11;26	0.000**
ABC-J stereotypic behavior	0	0;2.5	8	1;9.5	0.001**
ABC-J hyperactivity and	6	2;13	20	9.5;26.5	0.001**
ABC-J inappropriate speech	4	1;6.5	6	4.75;9.5	0.046*
FRPQ total	45	32.5;51	26	12;42.25	0.030*
FRPQ-P	12	7.5;13	6	2;10.75	0.049*
FRPQ-S	19	16;21	15	7.75;20	0.091
FRPQ-N	14	9;17.5	7.5	1.5;12.75	0.018*

Note. Typical, Probable, and Definite are the three categories participants were assigned to based on the standardized z-scores of the total SSP-J score, corresponding to z-scores above -1.00, between -1.00 and -2.00, and below -2.00, respectively. SSP-J = Short Sensory Profile, Japanese version; DEL = paternal deletion; mUPD = maternal uniparental disomy; PARS = Pervasive Developmental Disorders Autism Society Japan Rating Scale; ABC-J =

Aberrant Behavior Checklist, Japanese version; FRPQ = Food-Related Problem Questionnaire; FRPQ-P = Food-Related Problem Questionnaire – preoccupation with food; FRPQ-S = Food-Related Problem Questionnaire – impairment of satiety; FRPQ-N = Food-Related Problem Questionnaire – other food-related negative behaviors.

*$p < .05$. **$p < .01$