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Sensory Processing in Prader-Willi Syndrome

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Introduction

Prader-Willi syndrome (PWS) is characterized by considerable phenotypic complexities, some of which are aberrant behaviors, food related behaviors and, in some people, autistic-like behaviors that affect the quality of life of entire family systems.
As reported in the literature, people with PWS present with abnormalities in sensory responsiveness and sensory difficulties across behavioural, functional and quality-of-life outcomes *(Takahashi et al., 2019; Saima et al., 2022).* Nevertheless the relationship between sensory processing and autistic-like behaviors in people with PWS remains relatively unexplored (Saima et al., 2022).
This work aims at studying and identifying sensory processing dysfunction level by examining and comparing it with the severity of food-related behaviors and aberrant behaviors.

Materials and Methods

2.1. Study Design

A descriptive study was carried out. Three questionnaires were administered by clinical psychologists, working in the diagnostic services of Oasi Research Institute, throughout interviews with parents, as part of the psychological and psychoeducational assessment. Recruitment and organization of the sample are described in the following paragraph.

2.2 Participants

A total of 10 children with PWS, aged 30 to 144 months (7 males and 3 females; average chronological age 80,4 months, standard deviation 39.95), were consecutively recruited from specialized services of diagnosis and treatment of PWS during the year 2024.
All participants were diagnosed by a multidisciplinary team. One of them had normal intellectual functioning; four of them presented with comorbid Borderline Intellectual Functioning (BIF); two of them presented Mild Intellectual Disability (MID) and three of them had global developmental delay. Besides Five of them presented in comorbidity Language Developmental Disorders (LDD); two had Developmental Coordination Disorders; two had Attention Deficit/Hyperactivity Disorder (ADHD); two of them Distruptive Mood Dysregulation Disorder (DMDD).
Participants were divided into two subgroups: pre-scholar and scholar children.
The characteristics of the two subgroups are shown in Table 1.

Tab. 1: Characteristics of the sample

	Pre-scholar (n = 3) M ± SD	Scholar (n = 7) M ± SD
Age in months	38,33 ± 7,63	98.42 ± 33.34
IQ	71.33 ± 18.5	82,83 ± 12,12
Male/Female	2/1	5/2

2.3 Measures

A group of parents of pre-school and school age children completed the following check lists:

1. ***Social Responsiveness Scale, Second Edition*** (SRS-2; Constantino, 2012; D'Ardia et al., 2021) measures the presence and severity of social impairment of children from 2 years to 18 years old. It is mainly used with individuals with autism spectrum, family members, and others presenting with social impairments. It includes the following T-scores: Awareness (AWR); Cognition (COG); Motivation (MOT); Communication (COM); Restricted Interests and Repetitive Behavior (RRB); SRS Total; Social Communication and Interaction scale (SCIs); Restricted Interests and Repetitive Behavior scale (RRBs);
2. ***Sensory Processing Measure*** (SPM; Parham et al., 2007; Del Signore et al., 2020;) pre-scholar and scholar version to investigate sensory processing. The Home Form (75 items) requires just 15 to 20 minutes and includes the following T-scores: Social Participation, Vision, Hearing, Touch, Body Awareness (proprioception), Balance and Motion (vestibular function), Planning and Ideas (praxis), Total Sensory System;
3. ***Children's Eating Behavior Questionnaire*** (CEBQ; Wardle et al., 2001) is a tool for assessing children's eating styles. It is an interview including 35 items. A 5-point Likert scale is used, ranging from 1 = never to 5 = always. It includes eight scales: Food responsiveness, Emotional over-eating, Enjoyment of food, Desire to drink, Satiety responsiveness, Slowness in eating, Emotional under-eating, and Food fussiness. Higher scores correspond to a higher number of difficulties. For each scale there is a cut-off indicative of the presence/absence of atypia.

3. Statistical Analysis

Data analysis was conducted using the SPSS statistical package for Windows.
The means and standard deviations relating to the T scores of the tests applied are reported in Tables 2, 3 and 4 and in Graphics 2 A, 3 A e 4 A.
The results of the correlations carried out using Spearman's Rho test are reported in tables 5, 6 and 7.

4. Results

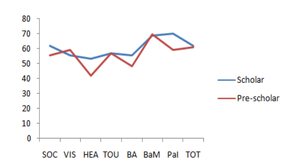
In the SPM test, both school-age and pre-school children achieved a total score in the "*Some difficulties*" range. The group of school-age children had overall higher scores than the preschool-age group. The latter had typical scores in the *Hearing* scale and *Body Awareness (proprioception)*. Both groups obtained scores in the "*Some difficulties*" range on the *Balance and Motion (vestibular function)* subscale, while the group of school-age children obtained a score in the "*Definite dysfunction*" range on the *Planning and Ideas (praxis)* scale. In the SRS-2 scale, the profiles of both groups are almost identical and are indicative of "*mild impairment*", with the exception of the *Restricted Interests and Repetitive Behavior* subscale which reflects a condition of "*moderate impairment*". Furthermore, the group of pre-school children is placed in a level of "moderate impairment" in the *Awareness* and *Motivation subscales.* The CEBQ profile reflects the characteristics of eating behavior typical of the syndrome, infact high scores are found for *Food responsiveness* and *Enjoyment of food*, as well as *Food fussiness.* While, the score of *Desire to drink* is low and this, as described in the literature, is a frequent characteristic of the syndrome.

Tab. 2: Means T scores of SPM

SPM	SOC	VIS	HEA	TOU	BA	BaM	Pal	TOT
Scholar	62	55,44	53,33	56,88	55,55	68,55	70	61,88
Pre-scholar	55,33	59	41,66	56,66	48	69,66	59,33	61

SOC = Social Participation; VIS = Vision; HEA = Hearing; TOU = Touch; BA = Body Awareness (proprioception); BaM = Balance and Motion (vestibular function); Pal = Planning and Ideas (praxis); TOT = Total Sensory System

Graph. 2: Means T scores of SPM



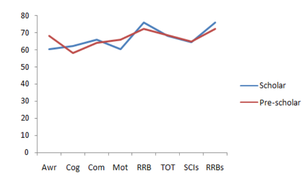
Definite dysfunction (T = 70-80); some difficulties (T= 60-69); typical (T = 40-59)

TAB. 3 Means T scores of SRS-2

SRS-2	Awr	Cog	Com	Mot	RRB	TOT	SCIs	RRBs
Scholar	60,22	62,11	65,88	60,22	75,88	68,22	64,44	75,88
Pre-scholar	68,33	58	64	66	72,33	68,66	64,66	72,33

AWR = Awareness; COG = Cognition; COM = Communication; MOT = Motivation; RRB = Restricted Interests and Repetitive Behavior; TOT = Total; SCI = Social Communication and Interaction scale; RRBs = Restricted Interests and Repetitive Behavior scale.

Graph. 3 A: Means T scores of SRS-2



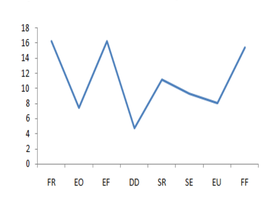
T-score of <59 = normal range; 60–65 = mild impairment; 66-75 = moderate impairment; T-score >75 = severe impairment.

TAB. 4 Means raw scores of CEBQ

CEBQ	FR	EO	EF	DD	SR	SE	EU	FF	TOT
	16,2	7,4	16,2	4,7	11,1	9,3	8	15,4	88,6

FR = Food responsiveness; EO = Emotional over-eating; EO = Enjoyment of food; DD = Desire to drink; SR = Satiety responsiveness; SE = Slowness in eating; EU = Emotional under-eating; FF = Food fussiness

Graph. 4 A: Means raw scores of CEBQ



The results of the Spearman's Rho test highlight significant correlations between the *Restricted Interests and Repetitive Behavior* and *Social Communication and Interaction* and *Social Participation* scales. *Hearing* is also related to the *Restricted Interests and Repetitive Behavior* scale. Besides, *Planning and Ideas (praxis)* are correlated to *Cognition* and the *Total score* with *Motivation*.
The scores reported above highlight a positive correlation between *proprioception* and *Emotional overeating*, the *Response to satiety* and the *Total score*. Furthermore, the *Response to food* and *Emotional overeating* are related to *planning/ideation*. Total scores on the CEBQ have a high correlation with *Body Awareness (proprioception)* and *Planning and Ideas (praxis)*. The total score of the CEBQ correlates with all the scales of the SPM test, except for *Awareness*.

Tab. 5 Correlations between SPM e SRS-2 (Spearman's Rho test)

	Awr	Cog	Com	Mot	RRB	TOT	SCIs	RRBs
SOC	-0,012	0,591	0,591	0,470	0,713*	0,606	0,632*	0,713*
VIS	-0,080	0,425	0,538	0,558	0,297	0,479	0,500	0,297
HEA	-0,393	0,578	0,572	0,437	0,673*	0,567	0,549	0,673*
TOU	0,495	0,171	0,573	0,848**	0,546	0,550	0,584	0,546
BA	0,467	0,099	0,301	0,157	0,143	0,313	0,226	0,143
BaM	0,380	0,073	0,233	0,233	0,167	0,292	0,211	0,167
Pal	-0,016	0,865**	0,504	0,390	0,625	0,512	0,571	0,625
TOT	0,275	0,488	0,543	0,665*	0,570	0,489	0,547	0,570

** p < 0,01; * < 0,05

Tab. 6 Correlation between SPM and CEBQ (Spearman's Rho test)

	FR	EO	EF	DD	SR	SE	EU	FF	TOT
SOC	-0,049	0,608	0,127	0,430	0,086	-0,136	0,247	0,064	0,621
VIS	0,402	-0,176	0,460	-0,144	-0,163	0,091	-0,468	-0,132	0,006
HEA	0,362	0,508	0,494	0,531	-0,166	-0,117	0,115	0,098	0,752*
TOU	0,061	0,265	0,409	0,606	-0,402	0,052	-0,037	0,509	0,722*
BA	0,451	0,690*	0,391	0,217	-0,652*	-0,247	-0,133	0,062	0,810**
BaM	0,247	0,551	0,296	-0,088	-0,391	-0,572	-0,217	-0,117	0,475
Pal	0,676*	0,669*	0,512	0,082	-0,550	-0,180	-0,234	-0,137	0,787**
TOT	0,278	0,574	0,449	0,300	-0,525	-0,344	-0,145	0,101	0,716*

** p < 0,01; * < 0,05

Tab. 7 Correlation between SRS-2 and CEBQ (Spearman's Rho test)

	FR	EO	EF	DD	SR	SE	EU	FF	TOT
Awr	-0,439	-0,056	-0,239	-0,185	0,092	-0,410	-0,130	0,486	-0,009
Cog	0,572	0,537	0,368	-0,045	-0,218	-0,071	-0,139	-0,126	0,642*
Com	0,199	0,182	0,526	0,516	-0,049	0,104	-0,034	0,420	0,676*
Mot	0,076	0,167	0,505	0,657*	-0,224	0,104	0,000	0,546	0,694*
RRB	0,187	0, 290	0,443	0,449	0,018	-0,058	-0,090	0,512	0,743*
TOT	0,187	0,176	0,497	0,502	0,018	0,117	-0,037	0,452	0,687*
SCI	0,226	0,289	0,469	0,477	-0,073	0,162	0,031	0,324	0,726*
RRB	0,187	0,290	0,443	0,449	0,018	-0,058	-0,090	0,512	0,743*

** p < 0,01; * < 0,05

SOC = Social Participation; VIS = Vision; HEA = Hearing; TOU = Touch; BA = Body Awareness (proprioception); BaM = Balance and Motion (vestibular function); Pal = Planning and Ideas (praxis); TOT = Total Sensory System; AWR = Awareness; COG = Cognition; COM = Communication; MOT = Motivation; RRB = Restricted Interests and Repetitive Behavior; TOT = Total; SCI = Social Communication and Interaction scale; RRBs = Restricted Interests and Repetitive Behavior scale; FR = Food responsiveness; EO = Emotional over-eating; EO = Enjoyment of food; DD = Desire to drink; SR = Satiety responsiveness; SE = Slowness in eating; EU = Emotional under-eating; FF = Food fussiness

Discussion and Conclusions

Children with SPW have "*some difficulties*" in sensory processing with particular reference to vestibular function and, furthermore, school age group have greater difficulties in planning and ideomotor skills. The results of the SRS-2 test confirm what has been reported in the literature regarding the presence of autistic-like behaviors, especially restricted and stereotyped interests. In pre-school children, there are also difficulties in awareness and motivation. The profile recorded in the CEBQ test also confirms some of the main characteristics of eating behavior found in the syndrome. Correlations between the instruments revealed that a higher level of difficulties in social participation is associated with the presence of restricted and stereotyped behaviors. Another interesting result that emerged is that deficits in proprioception are correlated with emotional overeating and a poor ability to perceive the sense of satiety. The correlations between the total of the CEBQ test and the different scales of the SRS-2 test, except awareness, reveal a relationship between autistic characteristics and eating problems.
The results obtained, although referring to a small sample, highlight the usefulness of including specific training for deficits in sensory processing and an intervention on autistic-type characteristics in the re-habilitation treatment of people with SPW.