Sensory Processing in Prader-Willi Syndrome

Serafino Buono*, Paola Occhipinti†, Marinnella Zingale*, Donatella Greco ** and Angela Antonia Costanzo***

*Unit of Psychology; ** Unit of Pediatrics; ***Unit of Pedagogy
Oasi Research Institute-IRCCS, Troina, Italy
E-mail: f.buono@oasi.en.it; pocchipinti@oasi.en.it; mzingale@oasi.en.it; dgreco@oasi.en.it; acostanzo@oasi.en.it

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., 2016; Saia et al., 2022). Nevertheless, the relationship between sensory processing and autistic-like behaviors in people with PWS remains relatively unexplored (Saia 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 3 to 14 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-schooler and school children.

The characteristics of the two subgroups are shown in Table 1.

2.3 Measures

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

- Sensory Processing Measure (SPM); Parham et al., 2007; Del Signore et al., 2020), pre-schooler 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, Body Awareness ( proprioception), Balance and Motion (vestibular function), Planning and Ideas (praxis), Total Sensory System;

- Children’s Eating Behavior Questionnaire (CEBQ; Wardle et al., 2001) is a tool for assessing children’s eating habits. 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 eating, Enjoyment of food, Desire to drink, Satisfaction 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 atypical behavior.

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 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 “Moderate Impairment” subscale of the “Moderate Differences” 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, in fact, high scores are found for Food responsiveness and Enjoyment of food. While, the score of Desire to drink is low and this, as described in the literature, is a frequent characteristic of the syndrome.

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 Eating representation. 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.

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.