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Introduction

Prader-Willi syndrome (PWS) is characterized by intellectual disability, learning disorders and behavioral problems, being required specific therapeutic interventions throughout their lives (Arnaiz-PaezBello-Ulloa, Román-García &, 2016, Franco,Stella, Beltrán & Bilbao Ramírez, 2019).

The extent of cognitive deficit can be variable, though is usually mild to moderate. The gold standard to measure cognitive impairment are Wechsler scales of intelligence for children and adults (Wais and Wisc III). However, most individuals with PWS have mild to moderate intellectual disability and concrete thinking thereby leading to failure in the interpretation of tasks and suboptimal executions. It is necessary to have evaluation tools that consider these difficulties, and also allow greater sensitivity to changes or improvements through treatment. For this reason, we develop an instrument that allows defining a Personalized Learning Profile (PLP) for people with PWS.

This instrument is being tested in our patients to expand knowledge regarding impairment of functions related to reading, writing, spatial temporal organization, management and recognition of emotions, visual-motor coordination and numerical calculation; regardless of age.

Aim

The purpose of this study was to assess the usefulness of a self-designed tool to identify strengths and weaknesses of cognitive characteristics in subjects with PWS.

Methods

This was a non-experimental, descriptive, cross-sectional study. The sample comprised 21 individuals with PWS of both sexes, between 10 to 40 years, who regularly attend transdisciplinary treatment at the SPINE Foundation.

In order to identify strengths and weaknesses in the cognitive profile, subjects were evaluated using the Wisc-III and Wais-III scales, as well as a novel instrument designed in our institution aimed at defining a personalized learning profile (PLP).

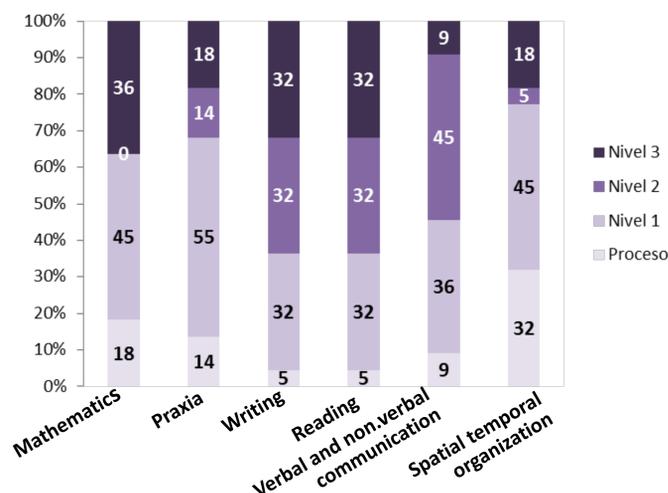
The department of Psychopedagogy of the Fundación SPINE works upon the development this profile, that evaluates the main difficulties and cognitive strengths. This PLP is obtained from a series of activities that are proposed to the subject, aimed at evaluating temporo-spatial organization, writing, reading, numbering, quantity notions, visual-motor coordination, text comprehension, and recognition of emotions. Each activity is classified as process or not achieved; and different levels (1-3) where the highest level (3) would indicate a strength in the area.

Results

According to the results of the Wisc-III and Wais-III scales, patients showed similar scores in verbal scale 60.2 ± 8.3 , execution scale 62.3 ± 11.1 and global IQ 57.6 ± 9.0 .

According to the data obtained with the designed instrument (PLP). Most of the sample showed strengths in:

- Mathematics, 36% reached level 3, which is related to numbering; a logical mathematical reasoning with greater abstraction.
- Reading and Writing, 32% reached level 3 in this areas. Which is linked to text writing, word dictation, free writing, search for hidden words. Showing greater phonological awareness.
- Only 18% of the subjects have passed tests related to visomotor coordination and human figure drawing (praxia).
- They have also showed some difficulties regarding temporal location and spatial notions, only 18% reached level 3 in this area.
- With respect to nonverbal language, empathy, expression of emotions and comprehension of texts, 9% have passed these tests.



Conclusions

The main cognitive difficulties were found in sequential information processing, working memory, logical-mathematical reasoning, the acceptance or understanding of others point of view, verbal auditory processing, attention, concentration, spontaneous and creative writing.

While the main strengths identified involved long-term memory and perceptual organization, sums of units with concrete material, writing and reading words.

We develop and demonstrated the usefulness of a Personalized Learning Profile (PLP) for people with PWS. With the gold standard measure (Wisc Wais) we couldn't be able to find a personalized profile, results only showed decreased performance in all areas. Whereas PLP measure allows to identify strengths and weaknesses in cognitive profile of people with PWS. This is of great relevance for the therapeutic approach, very helpful for designing specific intervention strategies aimed at rehabilitating impaired cognitive abilities.

References:

- Bello-Ulloa, L., Román-García, M., & Arnaiz-Paez, Y. (2016). Consideraciones históricas del Síndrome Prader-Willi; la atención educativa de este síndrome. *Educación y Sociedad*, 14(2), 27-40.
- Franco, M., Stella, D., La Rosa, D., Beltrán, E., & Bilbao Ramírez, J. L. (2019). Intervención multidisciplinaria en paciente diagnosticado con el síndrome de Prader-Will. *Revista Cubana de Pediatría*, 91(2).