

ORZI, M.; STEGMANN, J.

Fundación SPINE Socio.Psico.Inmuno.Neuro.Endocrinología | mercedesorzi@gmail.com

Introduction

Individuals with Prader Willi Syndrome (PWS) have specific characteristics such as muscular hypotonia, short height, hypogonadism, intellectual disability, psychomotor delay, behavioral and psychiatric difficulties. Regarding the difficulties related to speech and language manifestations, these individuals can show alterations in speech articulation, hyper or hyponasality, and limitations in receptive, expressive language and pragmatic abilities.

Aims

We aim to describe the linguistic profile of people with PWS, in particular of expressive and receptive language, and its relationship with the verbal intelligence quotient (VIQ).

Methods

The sample consisted on 12 individuals with SPW between 11 and 45 years old, without growth hormone therapy. We carried on a non-experimental, descriptive, cross-sectional study. All participants regularly attended to a transdisciplinary treatment at Fundación SPINE. The treatment consists on a comprehensive social-psycho-immuno-neuro-endocrinology approach. Clinical Evaluation of Language Fundamentals-4 (CELF-4) was used to assess linguistic profile involving semantic, morphology, syntactic and pragmatic aspects.

Results

Principals results showed a more favourable performance regarding word definitions (5.0±2.5), number repetition (4.5±3.5) and formulated sentences (3.8±2.0). On the other hand, a worse performance was found in understanding spoken paragraphs (2.4±4.5), recalling sentences (2.8±1.8) and word classes total (2.9±2.3). When evaluating the relationship between the CELF 4 subtests and VIQ, we only identified a significant relationship regarding the working memory subtest (r: 0.73, p= 0.01).

Table 1. CELF-4 scores

CELF-4 subtest	Score (mean±SD)
Recalling sentences	2.83±1.8
Formulated sentences	3.83±2.1
Word Classes Receptive	2.67±2.6
Word Classes Expressive	3.58±2.3
Word Classes Total	2.92±2.3
Word definitions	5.09±2.5
Understanding spoken paragraphs	2.42±4.3
Number repetition total	4.5±3.6
Familiar sequences 1Y2	3.92±3.8

Table 2. Relationship between CELF-4 and the verbal intelligence quotient (VIQ).

Domain Spearman's correlation (Rho) p value

Domain	Spearman's correlation (Rho)	p value
Core Language Score	0.38	0.25
Receptive Language	0.14	0.67
Expressive Language	0.01	0.99
Language content	0.39	0.23
Language memory	0.27	0.42
Working memory	0.73	0.01

Conclusions

In this study, the language profile of patients with PWS showed a better performance on expressive language skills, compared to comprehensive skills. We also identified an association between the VIQ and working memory. The main results were well bellow related with VIQ. Therefore, the VIQ is not a predictor of language skills but only of working memory.

Considering the complexity of these patients, It would be necessary for future studies to focus on pragmatic skills, speech, voice, and stomatognathic aspects to have a more complex profile.

References:

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