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Mental Health & Human Resilience International Journal
ISSN: 2578-5095

Neuropsychological and Emotional Aspects of Prader - Wiili Syndrome: A Brief Review

Dotta P*

Psychologist, University of Vale do Rio dos Sinos/UNISINOS, Brazil

***Corresponding author:** Patricia Dotta, Psychologist, Master in Clinical Psychology at the University of Vale do Rio dos Sinos/UNISINOS. Specialist in Neuropsychopedagogy and Neuropsychology, Teacher of Psychology at the Uniftec University Center in Bento-Gonçalves, Brazil, Email: patricia.dotta5@yahoo.com.br

Review Article

Volume 6 Issue 1

Received Date: March 14, 2022

Published Date: April 25, 2022

DOI: 10.23880/mhrij-16000170

Abstract

Prader-Willi Syndrome (PWS) is a complex multisystemic disorder characterized by several clinical manifestations, including intellectual deficiency, infant lethargy and hypotonia, and subsequent motor development impairment. The purpose of the review was to perform a brief review of literature on the genetic, neuropsychological and psychiatric aspects in Prader-Willi Syndrome. The present study is a review of exploratory literature in the online databases: LILACS, PubMed and Scielo. Patients with SPW predominantly have neuropsychological and psychological comorbidities associated with the syndrome. However, in Brazilian reality, studies focus on genetic aspects and management of problems associated with obesity. Psychological, neuropsychological and behavioral problems come second without proper intervention. The development of researches in Brazilian reality about neuropsychological and psychological aspects of such patients is suggested.

Keywords: Prader-Willi Syndrome; Obesity; Hypotonia; Neuropsychological Function; Bipolar Disorder

Introduction

Prader-Willi syndrome is a rare neurobehavioral disorder, characterized mainly by hypotonia, mental retardation, compulsive overeating and obesity connected to hypothalamic dysfunctions that can result in emotional, behavioral and neurological labilities [1,2]. For authors Noordam C, et al. [3] the cause of the syndrome is genetic, and it occurs in its large majority by the absence of the chromosome 15 inherited from the father. However, in some cases, it may be a maternal uniparental disomy, in which one inherits two copies of the mother's chromosome 15 and does not inherite something from the father and in rare cases can have a mistake in the process of "imprinting'" which leaves the inherited part of the father non-functional [4]. Such genetic error causes the main disorders in hypothalamus and in the pituitary gland which determines the symptoms of the syndrome, besides developing difficulties to regulate their emotions, and behavioral, relational and health problems [1,4].

SPW is characterized by a much wider neurodevelopment profile, including a distinct set of behavioral characteristics and mental retardation or learning difficulties. Even so, this profile is not universal or uniform; there is a considerable phenotypic variability observed among individuals with the syndrome [5,6]. Neuropsychological difficulties are described as a failure in the flexibility of executive functions, especially those associated with cognitive, communication and social skills [7,8]. The symptoms described above can also be found in patients with autistic spectrum disorders and lesional pathologies of the frontal lobe. In both cases:

patients with SPW and autists, dysfunctions in the frontal cognitive processes, like attention, work memory and executive functions [8,9].

It was reported that individuals with PWS have better visual-space performance and simultaneous processing tasks than verbal ones and those of sequential processing [9,10]; short-term memory is more severely affected than long-term memory [11-13]; auditory processing is poorer than visual processing [14] and the deficits were observed in fine motor [9,15] and executive tasks [16,17].

Besides, literature points that SPW patients present associated psychiatric comorbidities [18,19]. Among them are schizophrenia, bipolar disorder, and psychosis, but a lack of consensus on the probable psychiatric diagnosis is still observed [7,20]. However, a variety of clinical features has been described, including anxiety, agitation, states of confusion, persecutory delusions, disturbed sleep, acute onset without clear precipitating, mutant symptomatology, and a strong affective component [18]. Such patients can have compulsive behavior, presence of anxious symptoms, and dermatillomania are associated with the genetic alteration of the syndrome [18,20].

Severe behavioral problems are common and often present as stubbornness, challenge, easy frustrations, and fastness with anger. They usually have mood disorders, an important inability to regulate their emotions, obsessive behaviors, autistic traits, and have a great probability to unleash psychotic conditions at the end of adolescence or the beginning of adulthood [21,22]. The study conducted by Riznichuk MO, et al. [23]) indicates that often syndrome may be associated with opposing and aggressive behavior made to behavioral disorders and not necessarily to obesity and child lethargy.

In Brazilian literature, studies on PWS point to the importance of early diagnosis and the clinical characteristics of children [24]. However, the study of neuropsychological and psychotherapeutic aspects is scarce. Theoretical robustness in the international literature about behavioral and emotional problems in such patients is observed.

However, the look at patients with PWS and the knowledge beyond genetic and descriptive behavioral specificities, but of psychotherapeutic processes and results in psychotherapy and neuropsychological aspects is essential. It's believed it would be interesting to analyze the existence of deficits in cognitive processes related to the frontal lobe, like attention, memory, and executive functions in patients with PWS [25].

Clinical observations suggest that, regardless of genetic vulnerability, cognitive rigidity described as characteristic of the syndrome, combined with the difficulty of naming and expressing feelings and dealing with conflict, can predispose to an increase in the incidence of mood disorders, especially when a major neuropsychological loss is involved. The genetic component of the syndrome may be associated with psychic difficulties and environmental stress factors. Researches regarding this association are underway to comprehend in greater depth the emotional and neuropsychological specificities [9,18,25].

Conclusion

It was concluded that Prader-Willi Syndrome (PWS) occasions several demonstrations of physical and behavioral nature in individuals affected by it and about cognitive aspects, the scientific literature gives great emphasis on the intellectual disability in many of the diagnosed cases [26]. However, there is a great variety in the behavioral, psychological, and psychiatric manifestations that are little studied and perceived by the health professionals of Brazilian reality in the treatment of the patient. That suggests a difficulty in considering the child's singularity, psychological and neuropsychological function, even when it presents some genetic syndrome with previously considered specific characteristics. Besides, it's necessary that tools and training for an early diagnosis of this condition are promoted, to provide families with proper multi-professional care to insert these children into life in society. Psychoeducation, parental guidance, and medical and psychological supervision are key to the life quality of the child with the syndrome. Neuropsychological evaluation, in addition to identifying important points, has the role of promoting the search for the individual's life improvement, once it leads to the restitution of important decision making to establish a balance in front of the [27]. It is still necessary to perform more studies on such a theme to use more effective psychological and behavioral interventions that improve the patient's life.

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