

Papeles del Psicólogo (2022) 43 (2) 125-132

### Papeles del Psicólogo Psychologist Papers

https://www.papelesdelpsicologo.es/ • ISSN 0214-7823

Revista del Consejo General de la Psicología de España



Article

# West Syndrome and associated Autism Spectrum Disorder: Proposal for a neuropsychological assessment and intervention protocol

Christian López-Cruz<sup>1</sup>, Irene Cano-López<sup>1</sup>, Marta Aliño<sup>1</sup> and Sara Puig-Pérez<sup>1</sup>

<sup>1</sup> Universidad Internacional de Valencia.

#### ARTICLE INFO

#### Received: June 23, 2021 Accepted: December 10, 2021

# Keywords: West Syndrome Epilepsy Pediatric Epileptic Syndrome Autism Spectrum Disorder Neuropsychological Intervention

#### **ABSTRACT**

Patients with West Syndrome and associated Autism Spectrum Disorder (ASD) have cognitive deficits (i.e., attentional, mnestic, visuoperceptive, executive function, and language impairments) that may affect their quality of life. After delimiting the cognitive profile of these patients, this study aims to design a specific neuropsychological assessment and intervention protocol, from a holistic and integrative approach. The program consists of 48 sessions planned over six months, including neuropsychological assessment sessions (before, halfway through, and at the end of the intervention) and intervention sessions focused on the affected cognitive domains, socioemotional aspects, and the improvement of autonomy and functionality. The proposed program is expected to be effective in improving cognitive functioning and quality of life in this population, contributing to optimize health care.

## Síndrome de West y Trastorno del Espectro del Autismo asociado: Una propuesta de protocolo de evaluación e intervención neuropsicológica

#### RESUMEN

Palabras clave: Sindrome de West Epilepsia Síndrome Epiléptico Pediátrico Trastorno del Espectro del Autismo Intervención Neuropsicológica Los pacientes con Síndrome de West y Trastorno del Espectro del Autismo (TEA) asociado presentan déficits cognitivos (i.e., alteraciones atencionales, mnésicas, visuoperceptivas, en función ejecutiva y lenguaje) que pueden afectar a su calidad de vida. Tras delimitar el perfil cognitivo de estos pacientes, este estudio pretende diseñar un protocolo de evaluación e intervención neuropsicológica específico, desde un enfoque holístico e integrativo. El programa consta de 48 sesiones planificadas en seis meses, incluyendo sesiones de evaluación neuropsicológica (antes, a mitad y al final de la intervención) y sesiones de intervención focalizadas en los dominios cognitivos afectados, los aspectos socioemocionales y la mejora de la autonomía y funcionalidad. Se espera que el programa propuesto sea eficaz para mejorar el funcionamiento cognitivo y la calidad de vida de esta población, contribuyendo a optimizar la atención sanitaria.

#### Introduction

#### Pediatric epileptic syndromes

Epilepsy is a neurological disease that consists of a predisposition to suffer continuous epileptic seizures and associated neurobiological, cognitive, emotional, or social alterations (Fisher et al., 2005). It is a heterogeneous disease, which encompasses different electro-clinical syndromes, including West Syndrome (Besag, et al., 2016).

West syndrome is the most common pediatric encephalopathy, with an incidence of 1.6/100,000 persons (Arregui, et al., 2020). It begins at approximately four to seven months of age, and is characterized by infantile spasms (i.e., jerky, brief axial contractions that may be accompanied by crying, neck twitching, and nystagmus) and neurodevelopmental regression (Mytinger, 2021). Some patients also present with hypsarrhythmia (i.e., disorganized electroencephalographic pattern of bilateral, symmetrical slow waves without interhemispheric synchrony), although the diagnostic relevance of this indicator has recently been questioned (Mytinger, 2021). In 71-80% of cases it is associated with comorbid intellectual disability (Besag, 2006).

Both the seizures and the different treatments used to control them (i.e., antiepileptic drugs) can promote relatively variable cognitive deficits. This has led to an increased interest in indicating early neuropsychological assessment that is tailored to this population (Helmstaedter, et al., 2020; Vogt, et al., 2017). In this regard, the E-PILEPSY Consortium recommends placing special emphasis on the following domains: memory, attention, executive functions, language, visuospatial skills, intelligence, depression, anxiety, and quality of life (Vogt, et al., 2017).

#### West Syndrome and associated autism

West Syndrome has been associated with one of the most important neurodevelopmental disorders: autism spectrum disorder (ASD) (Besag, et al., 2016). If one assesses the great heterogeneity of symptoms that characterizes both conditions, this is not surprising. But, due to its severity, this comorbidity requires special attention.

Approximately 80% of patients with epilepsy or ASD present intellectual disability, which would explain why epilepsy has been associated more with intellectual disability than with autism (Legido, 2015). However, given that patients with ASD with a mild level of disability or exceptional abilities (i.e., savant) also present epileptic seizures, it is considered appropriate to speak of comorbidity (Muñoz et al., 2003). Thus, it has been found that approximately 20% of patients with West Syndrome present with associated ASD (Strasser, et al., 2018), suggesting that these clinical conditions share a common basis (Chong, et al., 2018).

In order to delimit the heterogeneous symptomatology presented by children with ASD, the main diagnostic criteria proposed by the DSM-5 (APA, 2013) are: difficulties in social interaction and communication, language delay and/or impairment, and the presence of stereotyped patterns, restrictive behavior, and particular interests. However, the autism spectrum encompasses many other domains, including processing speed, learning and verbal memory, reasoning, and problem solving (Velikonja, et al., 2019).

As for the cognitive and emotional alterations presented by children with West syndrome and associated ASD, they are not excessively different from those previously mentioned as sequelae of epilepsy, although the comorbidity of the two syndromes could increase the severity of these alterations. At the cognitive level, this population presents alterations in memory, learning, attention, visuoperception, language, and executive functions (Comeche & Vallejo, 2005). In turn, alterations in executive functions have been associated with poor social cognition (van den Berg, et al., 2021). At the emotional level, anxiety-depressive symptoms directly affect quality of life (Comeche & Vallejo, 2005).

#### Neuropsychological interventions used in the literature

The typical neuropsychological and emotional alterations in patients with West Syndrome and associated ASD produce a worsening of their quality of life (Vogt, et al., 2017). Therefore, the therapeutic focus should be, primarily, on improving the patient's autonomy and quality of life. To this end, intervention programs have been developed for patients with epilepsy or ASD separately, but, to our knowledge, there are no specific intervention protocols for patients with West syndrome and associated ASD.

Regarding intervention programs aimed at children with epilepsy, the computerized intervention FORAMENRehab stands out, focused on improving attention (Saard, et al., 2017). This intervention addresses the different attentional components according to the patient's needs and has been shown to be effective in this population with a minimum of 10 sessions, although longer application is recommended (Kaldoja, et al., 2015; Saard, et al., 2017). Another program focused on attention, in addition to executive functions, is the Rehabilitation of EXecutive Function and ATtention (EXAT), which has been shown to be effective in epilepsy (Rantanen et al., 2020). On the other hand, the Cogmed program, focused on working memory and applied by computer, has promising results in children with epilepsy (MacAllister, et al., 2014).

Shifting the focus to patients with ASD, we find the Unstuck and On Target (UOT) program, aimed at improving executive functions (i.e., cognitive flexibility, planning, and problem solving) with a contextualized implementation in the patient's environment (Kenworthy, et al., 2014). This program has been shown to be more effective than a social skills treatment for the improvement of executive functions (Kenworthy, et al., 2014). Also noteworthy is the Treatment and Education of Autistic and Related Communication Handicapped Children (TEACCH; Mesibov et al., 2005), which seeks to achieve maximum independence of the subjects, motivate them to explore and learn, improve intellectual functions and motor skills, reduce family stress, and promote school adaptation (Mesibov et al., 2005). Another noteworthy program is the DIR Model: Developmental Individual Difference Relationship, Floor-time (Greenspan & Wieder, 2007), a program that aims to develop emotional capacities, individuality, and the relationships of the subject with his/her environment, giving the infant a leading role in achieving the objectives through challenges and spontaneous games (Greenspan & Wieder, 2007). For its part, the computerized intervention Caribbean Quest, focused on attention and executive functions, has shown efficacy in improving visual working memory and selective attention (Macoun, et al.,

2021). Overall, the importance of non-computerized training focused on executive functions, especially when delivered from an ecological perspective, has been suggested to support children with ASD to tolerate frustration and interact using socially appropriate strategies (Pasqualotto, et al., 2021).

It should be noted that neuropsychological intervention should be closely linked to ongoing diagnoses (Hyman, et al., 2020), but, to our knowledge, the literature does not include studies that have applied a neuropsychological intervention program in people with West Syndrome and associated autism. Therefore, this work focuses on designing an intervention program for this population, considering globally the characteristics of the different interventions applied separately, enabling us to delimit future lines of research that wish to bring the two problems together. The program will be defined under a holistic and integrative orientation, focusing on the improvement of the affected cognitive domains, the socioemotional aspects, and the increase of autonomy and functionality.

#### Proposal of a neuropsychological intervention program

#### Program design

A neuropsychological intervention program is proposed for patients between the ages of 7 and 12 years with West Syndrome and associated ASD.

The intervention program will last six months (Figure 1). It will be structured with two one-hour individual sessions per week, for a total of 48 sessions, including 9 assessment sessions and 39 intervention sessions. Three neuropsychological evaluation periods will be carried out: an initial one (before the intervention), one halfway through the intervention, and one at the end of the intervention. In each of them the following domains will be assessed: general maturational and intellectual capacity, attention (sustained and selective), executive functions (working memory, inhibition, cognitive flexibility, planning, and problem solving), and social skills. Each of these periods will occupy three sessions, in order to avoid the effects of fatigue or bias due to the particular circumstances of a given day, for a total of nine evaluation sessions out of the 48 sessions that make up the program.

#### Neuropsychological Assessment Protocol

The intervention protocol is summarized in Table 1 and consists of the neuropsychological tests detailed below.

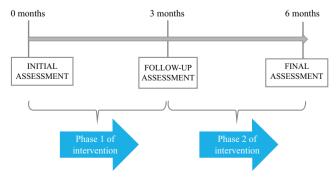


Figure 1. Intervention program design.

Table 1.

Cognitive processes assessed and tests used

Cognitive function	Process	Neuropsychological test
Intellectual functioning	Verbal comprehension Working memory Processing speed Fluid reasoning Visuospatial reasoning	Wechsler Intelligence Scale for Children (WISC-V; Wechsler, 2014)
Neuromaturational development	Language Visuoperception Executive functioning Memory Rhythm Laterality	Neuropsychological Maturity Questionnaire for Schoolchildren (CUMANES; Portellano et al., 2014)
Attention	Sustained attention Selective attention	Perception of Differences Test - Revised (FACES-R; Thurstone & Yela, 1979)
Executive functions	Cognitive flexibility Concept formation Sustained attentional capacity Inhibitory control	Wisconsin Card Sorting Test (WCST; Heaton et al., 1981) Stroop Color and Word Test (SCWT; Golden, 2001) Tower of Hanoi (Simon,
	Planning Working memory	1975)
Social skills	Autonomy Social adaptation Social intelligence	Psychosocial development assessment scale (Hurtig & Zazzo, 1971)

Wechsler Intelligence Scale (WISC-V; Wechsler, 2014). This scale assesses intellectual functioning. It contains numerous tests classified into five indices: verbal comprehension, working memory, processing speed, fluid reasoning, and visuospatial reasoning, and it provides an index of general intelligence functioning. It has a Cronbach's alpha ranging from 0.88 to 0.93 (Wechsler, 2014).

Cuestionario de Madurez neuropsicológica para escolares [Neuropsychological Maturity Questionnaire for schoolchildren] (CUMANES; Portellano, et al., 2014). This questionnaire assesses neuromaturational development, providing an index of global development resulting from the evaluation of 12 subscales grouped into six areas: language, visual perception, executive functioning, memory, rhythm, and laterality. It provides a profile that enables us to observe the weaknesses and strengths of each child. It has a Cronbach's alpha of 0.61-0.85 depending on the scale (Portellano, et al., 2014).

Difference perception test revisited (FACES-R; Thurstone & Yela, 1979). This test evaluates sustained and selective attention. It is composed of 60 graphic elements representing faces. It consists of determining which of the three faces is different and crossing it out in a maximum time of three minutes. A score of net correct answers (correct answers minus errors) and an index of impulsivity control is obtained. It has a Cronbach's alpha of 0.91 (Thurstone & Yela, 1979).

Wisconsin Card Sorting Test (WCST; Heaton, et al., 1981). This test assesses cognitive flexibility, concept formation, and sustained attentional capacity. It consists of 64 cards composed of shapes that vary in shape, color, and number of items. The individual is shown a card and is asked to match it with one of the four reference cards, without indicating the criteria to be followed. The evaluator only

indicates whether the matching is correct or not. After 10 correct trials, the classification principle changes without warning. The following indexes can be obtained: number of complete categories, number and percentage of perseverative errors, total number of errors, and "learning to learn" index. It has a Cronbach's alpha of 0.82-0.89 (Heaton, et al., 1981).

The Stroop Color and Word Test (Stroop; Golden, 2001). This test evaluates selective attention and inhibitory control. It consists of three sheets: a) word reading; b) color naming; and c) colorword test. It enables us to obtain three direct scores of the subject's performance on each sheet, and an interference index. It has a Cronbach's alpha of 0.85 (Golden, 2001).

The Tower of Hanoi (Simon, 1975). This test evaluates planning and working memory. It consists of a wooden platform with three vertical axes. The subject must be able to transfer the disks from the first to the third axis leaving them in the same initial position. It is specified that a large disk cannot be placed on top of a smaller one and no more than one disk can be moved at a time. The test is evaluated taking into account the number of movements made, the number of errors and the time taken to reach the target movement. It has a Cronbach's alpha of 0.71 (Humes, et al., 1997).

Echelle de développement psycho-social [Psychosocial development assessment scale] (Hurtig & Zazzo, 1971). This scale assesses autonomy, social adaptation, and social intelligence. It is composed of 139 items that evaluate three aspects: the acquisition of self-direction, to assess the level of autonomy; the development of interests on social aspects, reading, and intellectual openness; and the development of interpersonal relationships, to assess dependence on parents and peers. It enables us to obtain a development coefficient. It has a Cronbach's alpha of between 0.66 and 0.96 (Aranda-Sánchez, et al, 1995).

#### Neuropsychological Intervention Protocol

A total of 39 sessions will be devoted to the neuropsychological intervention itself. These sessions will consist of the activities described below, interspersed according to criteria of variety and complexity. As a general rule, the emotional component will always be present and will progress from attentional capacity to executive functions to more complex domains such as social skills or problem solving. It should be noted that it is of interest for patients to become familiar with the tests in order to improve their perception of self-efficacy and to accentuate the daily evolution, so an efficient balance between novelty and practice must be maintained. On the other hand, the duration of the sessions will allow them to contain three different exercises of approximately fifteen to twenty minutes each. A description of the tasks is given below.

Letter and number matrix. This is a pencil and paper task for the improvement of sustained attention and planning. It comprises a grid containing ten symbols (five vowels and five numbers) repeated and placed in a disordered way. It consists of making a circle around each letter or number using different colors and without leaving any out. Then, the total number of times that one of these symbols appears is counted and the possible mistakes are reviewed. The difficulty can be increased by replacing the letters and numbers with symbols or by increasing the size of the matrix.

Pattern search. This consists of a task for the improvement of cognitive flexibility. For this purpose, a deck of cards should be

used, and a pile should be placed face down. The patient is asked to uncover the cards one at a time and place them face up on the table. The objective is for him/her to discover the pattern that the cards should follow, without having been given any prior instruction. Some examples of hidden rules include ordering the cards according to a numerical criterion in an increasing or decreasing direction or according to color. As the patient discovers the hidden criterion, feedback will be given. In the case of the second answer, the selected card will be returned to the pile. The therapist may, at any time and without prior warning, change the criterion, forcing the patient to make his/her answer more flexible in order to get it right again.

Sudokus. This is an exercise to improve selective attention, working memory, and planning. The patient must fill in, with numbers from one to nine, a sheet of paper with a printed grid with some blank spaces and others with the numbers already prefixed, without repeating any of the squares, columns, or lines. The complexity of the Sudoku may vary to suit the individual case, and guidelines will be given to facilitate its completion.

Problem Solving. This consists of a pencil-and-paper task to improve problem solving and social skills. A list of social, family, or economic problems is presented. First, the patient must write in his or her own words what he or she has understood about the problem presented. Secondly, he/she should propose three qualitatively different possible alternative solutions. Third, he/she must write down the pros and cons of each of the solutions. Finally, he/she is given feedback on their work and encouraged to change those aspects that could be improved.

Copying a figure. This consists of a pencil and paper exercise to improve selective attention, working memory, and impulsivity. It consists of a sheet of paper with two grids formed by rows and columns filled with dots. In one of the grids, a figure is drawn following the union of certain points. On the other, the patient must copy the drawing, paying attention to the correct row and column.

Scene identification. This is an exercise for the improvement of skills and interpretation of social scenes and emotional understanding. A scene from everyday life is presented on a sheet of paper representing a given social situation with several characters involved. The patient will have to identify three different elements in it: a) describe what has happened in the scene; b) indicate what each of the characters involved is thinking; c) describe the feelings of each character as a consequence of the situation. After each interpretation, feedback will be provided.

How I feel. This consists of a task aimed at improving the identification of emotions and the management of frustration in the daily context. It consists of an illustrated sheet with faces representing certain emotions, with varying complexity (from basic emotions to more complex ones). For each emotion, the patient must describe a situation in which he/she has felt this way since the last session.

Clear language. This is a dynamic task that increases the capacity for social interaction and the use of appropriate language and conversational skills. It consists of a conversation on a topic of the patient's own choice in which the person who is holding a pen representing a symbolic microphone will have the turn to speak. In this way, the patient can only speak when he/she has the pen in his/her possession. The aim of the task is to teach the patient to respectfully take turns to speak, to keep their gaze on the

person they are talking to, and to show interest in the other person's concerns, avoiding egocentrism and perseveration.

#### Description of program sessions

Figure 2 shows a summary schedule of the program sessions, which are detailed below. In each session, five minutes will be reserved for the patient to take a break.

Sessions 1-3. The baseline neuropsychological assessment is performed.

Session 4. This is aimed at attention training. The session begins with the activity "How I feel". The duration is 15 minutes and, throughout the exercise, the therapist should ask the patient about the situations described that seem most relevant, in addition to giving feedback on their emotional understanding. After this first exercise, the tasks of "Letters and numbers matrix" (20 minutes) and "Sudokus" (20 minutes) are performed.

Session 5. This session is dedicated to the intervention of attentional capacity. It will start with the "How I feel" task (15 minutes). This will be followed by the "Sudoku" (20 minutes) and the "Copying a figure" task (20 minutes). In this third exercise of the session, in addition to the work involved in the previous task, the prevention of impulsive behavior will be added.

Session 6. This session is devoted to the training of attention, both sustained and selective. It will start with the "How I feel" task (15 minutes). Subsequently, the "Copying a figure" task (20 minutes) and the "Letters and numbers matrix " task (20 minutes) will be performed.

Sessions 7-13. These sessions follow the same sequence as the three previous sessions, with the possibility of changing the order of the attention exercises, in order to give more variety to the sessions. Likewise, materials adapted to the needs and preferences of each patient will be used. Once the attentional capacity has been addressed, in the following sessions exercises with new elements and domains on which to intervene will be introduced.

Session 14. This session is intended to introduce executive function training. The session will start with the "How I feel" task (15 minutes). Then, it will proceed with the "Letters and numbers matrix" task (20 minutes), and it will end with the "Search for the pattern" activity (20 minutes).

Session 15. Continuing with the introduction of executive function training, this session will continue to present new exercises. We will start with the "How I feel" task (15 minutes). This will be followed by a "Sudoku" (20 minutes), and the "Problem solving" task (20 minutes) will be introduced.

Session 16. This session continues with the improvement of attention and social skills, but also emphasizes emotional understanding. It will start with the "How I feel" task (15 minutes). This will be followed by "Copying a figure" (20 minutes) and will end with the "Identifying scenes" activity (20 minutes).

Session 17. This is the last session in which an unfamiliar exercise will be introduced, aimed at continuing to improve social skills and emotional understanding. It will start with the usual "How I feel" task (15 minutes), followed by "Letters and numbers matrix" (20 minutes) and the novel "Clear language" task (20 minutes).

Sessions 18-36. Sessions 18-24 and 28-36 will follow the same sequence as the previous four sessions and will be separated by the follow-up neuropsychological assessment (sessions 25-27). In this second set of sessions a differentiation is made between two blocks of exercises: those aimed at improving attention and executive functions (Letters and numbers matrix, Sudoku, and Copying a figure), and those aimed at social skills, emotional understanding, and cognitive flexibility (pattern search, problem solving, scene identification, and clear language). The continuity of the present structure, one activity per block, in addition to the initial "How I feel", is required for the realization of future sessions. Likewise, the sequence is required to be the same, performing the attention task first and the social skills task second. However, since the number of tasks in both blocks is unclear, the alternation of exercises between blocks is urged, such that a wheel is made that allows for all possible combinations.

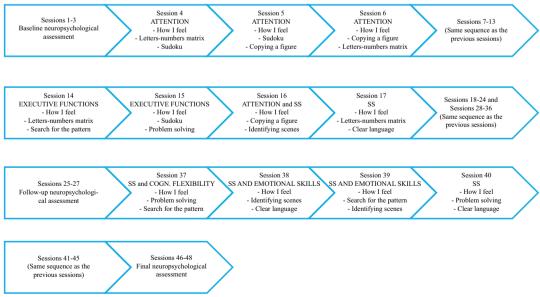


Figure 2. Summary schedule of sessions.

Note: SS: social skills.

Once this interval of sessions has concluded, it is assumed that the attention and executive functions of the first block have been practiced satisfactorily and a last period in which only the tasks of the second block are used. This is carried out as follows.

Session 37. This session is aimed at training in social skills and cognitive flexibility. It will start with the "How I feel" task (15 minutes). Then, 20 minutes will be devoted to "Problem solving" and the last 20 minutes for "Searching for the pattern".

Session 38. Emphasizing the development of social skills and understanding of emotions, this session will begin with "How I feel" (15 min), continue with the task of "Identifying scenes" (20 min), and end with "Clear language" (20 min).

Session 39. Continuing with the same focus as the previous two sessions, this session will begin with "How I feel" (15 minutes) and continue with "Searching for the pattern" (20 minutes), ending with "Identifying scenes" (20 minutes).

Session 40. In this session we will work again with special emphasis on social skills. After the usual 15 minutes of "How I feel", the "Problem solving" task (20 minutes) will be followed by the "Clear language" task (20 minutes).

Sessions 41-45. These will follow the same layout and sequence as the previous four sessions, with the possibility of changing the order of the exercises proposed per session, in order to give more variety to the sessions.

Sessions 46-48. The final neuropsychological evaluation will be performed.

#### Conclusions

This protocol is intended to become a rehabilitation model for children with West Syndrome and associated ASD. For this purpose, a working methodology focused on the improvement of cognitive, socioemotional, autonomy, and contextual functionality aspects has been established. To this end, a program of 48 sessions to be implemented in a six-month interval is presented, including intervention sessions and evaluation sessions aimed at assessing the effectiveness of the program.

The cognitive alterations that can be found as a consequence of suffering from a pediatric epileptic syndrome and associated autism are diverse. For this reason, it has been proposed to evaluate and intervene in domains such as attention, memory, executive functions, language, visuospatial skills, mood, and quality of life (Helmstaedter, et al., 2020; Vogt, et al., 2017). For the design of this intervention, all these domains have been taken into account, creating a comprehensive rehabilitation process. In view of this approach, it seems consistent to expect an improvement in general intellectual ability, estimated by the WISC-V general intelligence functioning index (Wechsler, 2014). Likewise, of the four subindices that make up this test, an increase in working memory and processing speed scores is hypothesized due to the special emphasis placed on the domains of attention and executive functions. Similarly, it is expected that an improvement would be found in the global development index of the CUMANES test (Portellano, et al., 2014), which would indicate a general increase in neuromaturational development on the test.

Going into detail on the specific aspects that are intended to be satisfied with this intervention, it is worth mentioning the functioning of attention, both selective and sustained in the short and long term. There are many programs that focus on this field, although in different populations. The FORAMENRehab program developed by Saard, et al. (2017) focused on the various processes of attention in an individualized intervention and with a minimum of 10 sessions for patients with epilepsy. The same was true for the EXAT program for patients with epilepsy, who had promising results in this field (Rantanen, et al., 2020). Considering these precedents, an improvement in performance on the FACES-R test (Thurstone & Yela, 1979), the WSCT test (Heaton, et al., 1981), or the Stroop test (Golden, 2001) could be expected after the application of the proposed intervention program.

On the other hand, another aspect on which the proposed intervention has focused is the executive functions. Considering the previous literature, the proposed intervention is expected to improve executive functions (MacAllister, et al., 2014; Kenworthy, et al., 2014). Specifically, one would expect to find improvements in the impulsivity control index of the FACES-R test (Thurstone & Yela, 1979) or an increase in cognitive flexibility and concept formation ability in the WSCT test (Heaton, et al., 1981). Following the same logic, we expect to find a decrease in the interference index of the Stroop test (Golden, 2001), which assesses inhibitory control, and better performance in the Tower of Hanoi task (Simon, 1975), reflecting planning ability and working memory.

Finally, regarding the approach used for the design of the intervention and its domains, it is worth mentioning the socioemotional aspect, on which it has also been proposed to work. It is expected to obtain an improvement in the development coefficient of the scale for the evaluation of psychosocial development, involved in aspects such as autonomy, social adaptation, and social intelligence. Such an improvement can be expected considering that the program establishes specific tasks for this purpose, as well as the results found by Rantanen et al (2020), which showed improvements at the attentional and behavioral level after the application of the EXAT program in people with epilepsy. In fact, a recent systematic review shows that performance in executive functions is positively associated with social skills (van den Berg, et al., 2021).

There are several limitations to the intervention. On the one hand, most of the exercises proposed are in a clinical and penciland-paper context. This is not a negative thing per se, but it could make it difficult to extrapolate to everyday life. It is not that this aspect has been neglected. The emotional tasks applied in each session and the socioemotional tasks of certain specific sessions seek, precisely, to prepare the child to better cope in his/her daily context. However, the importance of working first in the more purely cognitive framework of attention and executive functions leads to a mechanics that may be more artificial. On the other hand, and due to the same aspects described above, the intervention may run the risk of becoming monotonous. The repetition of certain exercises, even if they are presented with a search for projection and improvement in their resolution and a continuous adaptation of the task, can lead to this problem. It will be down to the professional responsible for the intervention to create dynamism and flexibility in the session, attending to the particular needs of each patient in each circumstance, and thus improving their motivation in the rehabilitation process.

Despite these possible limitations, this intervention proposal focuses on a population that, while due to its infant nature presents a great projection of improvement for the present difficulties, requires more attention than the scientific community has given it so far. We are referring to the comorbidity of two severe pathologies: West Syndrome and ASD. The regressive nature of the aforementioned pathologies results in a heterogeneity of deficits that can encompass any neurodevelopmental domain, which justifies the recent recommendation for intervention in the pre-symptomatic period in children at risk of developing ASD (Grzadzinski, et al., 2021). Considering this, in the present work an intervention program has been designed based on the available scientific evidence, which aims to improve the functionality and quality of life of this population.

#### **Conflict of interest**

There is no conflict of interest.

#### References

- American Psychiatric Association (APA) (2013). Manual de diagnóstico y estadístico de los trastornos mentales, DSM 5. [Diagnostic and Statistical Manual of Mental Disorders, DSM 5], Editorial Médica Panamericana.
- Aranda-Sánchez, M., Brante Lara, M. E., & Robles Cárdenas, L. M. (1995). Estandarización de la escala de desarrollo psicosocial de René Zazzó y Marie Claude Hurtig en niños de cinco años de edad [Standardization of René Zazzó and Marie Claude Hurtig's psychosocial development scale in five-year-old children]. (PhD thesis, Universidad de Chile). http://repositorio.uchile.cl/handle/2250/134312
- Arregui, M. J. B., Prat, N. T., Salas, L. V., & Combalía, R. A. (2020). El síndrome de West o espasmos infantiles: Síntomas, etiología y opciones de tratamiento [West syndrome or infantile spasms: Symptoms, etiology, and treatment options]. Revista Sanitaria de Investigación, 1, 5.
- Besag, F. M. (2006). Cognitive and behavioral outcomes of epileptic syndromes: Implications for education and clinical practice. *Epilepsia*, 47, 119-125. https://doi.org/10.1111/j.1528-1167.2006.00709.x
- Besag, F., Gobbi, G., Aldenkamp, A., Caplan, R., Dunn, D. W., & Sillanpää, M. (2016). Psychiatric and behavioural disorders in children with epilepsy (ILAE Task Force Report): Behavioural and psychiatric disorders associated with epilepsy syndromes. *Epileptic Disorders*, 18, S37-S48. https://doi.org/10.1684/epd.2016.0815
- Chong, P. F., Saitsu, H., Sakai, Y., Imagi, T., Nakamura, R., Matsukura, M., Matsumoto, N., & Kira, R. (2018). Deletions of SCN2A and SCN3A genes in a patient with West syndrome and autistic spectrum disorder. *Seizure*, 60, 91-93. https://doi.org/10.1016/j.seizure.2018.06.012
- Comeche, M. & Vallejo, M.A. (2005). Manual de terapia de conducta en la infancia [Handbook of behavior therapy in childhood]. Dykinson.
- Fisher, R. S., Boas, W. V. E., Blume, W., Elger, C., Genton, P., Lee, P., & Engel Jr, J. (2005). Epileptic seizures and epilepsy: Definitions proposed by the International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE). *Epilepsia*, 46, 470-472. http://dx.doi.org/10.1111/j.0013-9580.2005.66104.x
- Golden C.J. (2001). Stroop: Test de colores y palabras [Stroop Color and Word Test]. TEA Ediciones.
- Greenspan, S. I., & Wieder, S. (2007). The developmental individual-difference, relationship- based (DIR/Floortime) model approach to autism spectrum disorders. In E. Hollander & E. Anagnostou (Eds.),

- Clinical manual for the treatment of autism (p. 179–209). American Psychiatric Publishing, Inc.
- Grzadzinski, R., Amso, D., Landa, R., Watson, L., Guralnick, M., Zwaigenbaum, L., Deák, G., Estes, A., Brian, J., Bath, K., Elison, J., Abbeduto, L., Wolff, J., & Piven, J. (2021). Pre-symptomatic intervention for autism spectrum disorder (ASD): Defining a research agenda. *Journal of Neurodevelopmental Disorders*, 13(1), 49. https://doi.org/10.1186/s11689-021-09393-y
- Heaton, R.K., Chelune. G.J., Talley, J.L., Kay, G.G. & Curtiss, G. (1981).
  Test de Clasificación de Tarjetas de Wisconsin [Wisconsin Card Sorting Test]. TEA Ediciones.
- Helmstaedter, C., Sadat-Hossieny, Z., Kanner, A. M., & Meador, K. J. (2020). Cognitive disorders in epilepsy II: Clinical targets, indications and selection of test instruments. *Seizure*, 83, 223-231. https://doi.org/10.1016/j.seizure.2020.09.031
- Humes, G. E., Welsh, M. C., Retzlaff, P., & Cookson, N. (1997). Towers of Hanoi and London: reliability and validity of two executive function tasks. *Assessment*, 4, 249-257.

http://dx.doi.org/10.1177/107319119700400305

- Hurtig, M.C. & Zazzo, R. (1971). La medida del desarrollo psicosocial [The measurement of psychosocial development]. In R. Zazzo: Manual para el examen psicológico del niño. Volumen II [Manual for the psychological examination of the child. Volume II]. Fundamentos Editorial.
- Hyman, S. L., Levy, S. E., Myers, S. M. (2020). Identification, evaluation, and management of children with autism spectrum disorder. *Pediatrics*, 145(1), e20193447. https://doi.org/10.1542/peds.2019-3447
- Kaldoja, M. L., Saard, M., Lange, K., Raud, T., Teeveer, O. K., & Kolk, A. (2015). Neuropsychological benefits of computer-assisted cognitive rehabilitation (using FORAMENRehab program) in children with mild traumatic brain injury or partial epilepsy: A pilot study. *Journal of Pediatric Rehabilitation Medicine*, 8, 271-283.

http://dx.doi.org/10.3233/PRM-150346

- Kenworthy, L., Anthony, L.G., Naiman, D.Q., Cannon, L., Wills, M.C., Luong-Tran, C., Werner, M.A., Alexander, K.C., Strang, J., Bal, E., & Sokoloff, J.L. (2014). Randomized controlled effectiveness trial of executive function intervention for children on the autism spectrum. *Journal of Child Psychology and Psychiatry*, 55, 374-383. http://dx.doi.org/10.1111/jcpp.12161
- Legido, A. (2015). Síndromes epilépticos pediátricos y autismo [Pediatric epileptic syndromes and autism]. Fundación Síndrome de West.
- MacAllister, W. S., Vasserman, M., Rosenthal, J., & Sherman, E. (2014). Attention and executive functions in children with epilepsy: what, why, and what to do. *Applied Neuropsychology: Child, 3*, 215-225. http://dx.doi.org/10.1080/21622965.2013.839605
- Macoun, S. J., Schneider, I., Bedir, B., Sheehan, J., & Sung, A. (2021).
  Pilot study of an attention and executive function cognitive intervention in children with Autism Spectrum Disorders. *Journal of Autism and Developmental Disorders*, 51(8), 2600-2610.
  https://doi.org/10.1007/s10803-020-04723-w
- Mesibov, G. B., Shea, V., & Schopler, E. (2005). The TEACCH approach to autism spectrum disorders. Springer Science & Business Media.
- Muñoz, J.M., Salvadó, B., Ortiz Alonso, T., Amo, C., Fernández Lucas, A., Maestú, F., & Palau Baduell, M. (2003). Clínica de la epilepsia en los trastornos del espectro autista [Epilepsy clinic in autism spectrum disorders]. Revista de Neurologia, 36, S61-S67. https://doi.org/10.33588/rn.36S1.2003075
- Mytinger, J. R. (2021). Definitions and diagnostic criteria for infantile spasms and West syndrome- historical perspectives and practical

- considerations. *Seminars in Pediatric Neurology, 38*, 100893. https://doi.org/10.1016/j.spen.2021.100893
- Pasqualotto, A., Mazzoni, N., Bentenuto, A., Mulè, A., Benso, F., & Venuti, P. (2021). Effects of cognitive training programs on executive function in children and adolescents with Autism Spectrum Disorder: A systematic review. *Brain Sciences*, 11(10), 1280. https://doi.org/10.3390/brainsci11101280
- Portellano, J. A. & García, J. (2014). Neuropsicología de la atención, las funciones ejecutivas y memoria [Neuropsychology of attention, executive functions and memory]. Síntesis.
- Rantanen, K., Vierikko, E., Eriksson, K., & Nieminen, P. (2020). Neuropsychological group rehabilitation on neurobehavioral comorbidities in children with epilepsy. *Epilepsy & Behavior*, 103, 106386. http://dx.doi.org/10.1016/j.yebeh.2019.06.030
- Saard, M., Kaldoja, M. L., Bachmann, M., Pertens, L., & Kolk, A. (2017). Neurorehabilitation with FORAMENRehab for attention impairment in children with epilepsy. *Epilepsy & Behavior*, 67, 111-121. http://dx.doi.org/10.1016/j.yebeh.2016.12.030
- Simon, H. A. (1975). The functional equivalence of problem solving skills. Cognitive Psychology, 7, 268-288. https://doi.org/10.1016/S0747-5632(03)00002-5

- Strasser, L., Downes, M., Kung, J., Cross, J. H., & De Haan, M. (2018).

  Prevalence and risk factors for autism spectrum disorder in epilepsy:

  A systematic review and meta-analysis. *Developmental Medicine and Child Neurology*, 60, 19-29. https://doi.org/10.1111/dmcn.13598
- Thurstone & M. Yela. (1979). CARAS-R. Test de Percepción de Diferencias-Revisado [FACES-R-Perception of differences test]. TEA Ediciones.
- van den Berg, L., de Weerd, A. W., Reuvekamp, M., & van der Meere, J. J. (2021). Associating executive dysfunction with behavioral and socioemotional problems in children with epilepsy. A systematic review. *Child Neuropsychology*, 27(5), 661-708. https://doi.org/10.1080/09297049.2021.1888906
- Velikonja, T., Fett, A. K., & Velthorst, E. (2019). Patterns of nonsocial and social cognitive functioning in adults with Autism Spectrum Disorder: A systematic review and meta-analysis. *JAMA Psychiatry*, 76(2), 135-151. https://doi.org/10.1001/jamapsychiatry.2018.3645
- Vogt, V.L., Äikiä, M., Del Barrio, A., Boon, P., Borbély, C., Bran, E., Braun, K., Carette, E., Clark, M., Cross, J.H., & Dimova, P. (2017). Current standards of neuropsychological assessment in epilepsy surgery centers across Europe. *Epilepsia [Epilepsy]*, 58, 343-355. http://dx.doi.org/10.1111/epi.13646
- Wechsler, D. (2014). WISC-V. Escala de Inteligencia de Wechsler para niños-V [Wechsler Intelligence Scale for Children]. Pearson Educación.