

Challenges in Sensory Integration and Processing in the Child with Wolf-Hirschhorn Syndrome

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ABSTRACT

Challenges in sensory integration and processing occur when the central nervous system is not able to adequately detect, interpret and respond to sensory information captured by the various sense organs. Difficulty integrating and processing sensory information adequately, impacts environmental interaction and the capacity to respond effectively to external stimuli and sensory input from one's own body. The present study's aim was to carry out a preliminary exploration of challenges in sensory integration and processing in children with Wolf-Hirschhorn Syndrome (WHS). Parents whose children (n=16) were between 3 and 10 years old completed a Short Sensory Profile. The most prominent areas of difficulty were obtained in the Low Energy / Weak and Under-Responsive / Seeks sensation sections of the Short Sensory Profile. The present study offers a preliminary view on the presence of challenges in sensory integration and processing in children with WHS.

INTRODUCTION

Dr. Jean Ayres, an American occupational therapist, was the first to describe a set of atypical behaviours related to poor sensory integration and processing [1-3]. She referred to this as a sensory integration dysfunction and described a situation in which the Central Nervous System (CNS) is not able to adequately detect, interpret and respond functionally to sensory information captured by the various sense organs [4]. She proposed that the way in which the CNS processes, integrates, and responds to sensation affects cognitive, motor, emotional, regulatory, and adaptive behaviour. Although the exact neuronal mechanisms related to sensory integration are not completely understood [5-7], the brain-behavior interactions posited by Ayres are gaining support and validation [4]. Challenges in sensory integration and processing include concerns about sensory reactivity (i.e., the process of modulating neuronal activity in response to sensory stimuli) and/or perception (i.e., the ability to recognize and interpret sensory stimuli). Sensory integration and processing issues can profoundly affect development and the capacity to participate in daily life occupations [4,8]. Simple activities such as eating, dressing, bathing or playing can become difficult challenges to overcome, and in many cases, the causes of the difficulty are attributed to other reasons. Challenges in sensory integration and processing are frequently associated with neurodevelopmental and genetic conditions [9-12]. It is estimated that 40% to 88% of children with a diagnosed disability experience challenges in sensory integration and processing [13-15]. However, little is known about the sensory concerns in people with Wolf-Hirschhorn Syndrome (WHS).

Wolf and Hirschhorn [16,17] described a genetic syndrome caused by a partial deletion of the short arm of chromosome 4 for the first time in 1965. Delays in motor functions, cognition, regulatory functions such as sleep-wake cycles and communication abilities are all reported to be highly prevalent in children with Wolf-Hirschhorn Syndrome (WHS) [18]. These types of functional difficulties have been linked with challenges in sensory integration and processing in other diagnostic groups [4]. Furthermore, occupational therapy clinical experience with children with WHS has raised concerns relative to difficulties processing sensory input. A common participation challenge, which motivates referrals to occupational therapy, is the lack of purposeful interaction with objects, which impacts play and self-care. For example, children with WHS often do not show interest in manipulating toys or using a spoon for self-feeding. The vestibular and proprioceptive systems are of particular concern, given that issues with posture and movement are often identified as factors affecting participation; poor posture and stability impact hand use for object manipulation. Perception of tactile input is also of concern given its role in learning new motor skills [19]. The present study's aim was to carry out a preliminary exploration of sensory integration issues in children with WHS.

METHODOLOGY

During the annual meeting (2016) of the Spanish Association of WHS (AESWH; abbreviation in Spanish), parents of three to ten-year-old children diagnosed with WHS were invited to complete the Short Sensory Profile [20]. This questionnaire is used to obtain information about children's reactions to everyday sensory experiences and screens for challenges in sensory integration and processing. The Short Sensory Profile (SSP) [20] is a 38 item condensed version of the original 125 item Sensory Profile (SP) [21]. The validity of these tools is well established and sensory questionnaires are considered an acceptable method of screening for challenges in sensory processing [21,22]. The SSP, translated to Spanish (S-SSP) for use with Spanish speakers living in the United States and distributed by the publisher, has been revised and culturally adapted for Spain [23]. This revised version was used in the present study.

The 38 items of the SSP are divided into 7 categories: Tactile Sensitivity (8 items), Taste/Smell Sensitivity (4 items), Movement

Sensitivity (3 items), Underresponsive/Seeks Sensation (7 items), Auditory Filtering (6 items), Low Energy/Weak (6 items) and Visual/Auditory Sensitivity (5 items). The Sensitivity sections include items designed to capture over-reactivity to sensations. For example, refusal or expressions of fear or pain to sensory-laden activities are hypothesized to be manifestations of sensory over-reactivity [2,22,24-26]. The Auditory Filtering section includes items related to processing of auditory information, especially as it relates to using and screening out auditory input in daily life [21]. The items of the Underresponsive/Seeks Sensation section refer to behaviours linked to awareness of sensory stimuli in different sensory systems. For example, items such as not noticing that one's clothes are twisted or seeking intense movement are part of this section [21]. Finally, the items of the Low Energy/Weak section reflect behaviours related to the ability to use muscles and move [21]. Difficulties in this area can be manifestations of challenges processing proprioceptive input [19] and items refer to observations such as weak muscles and postural difficulties [21].

Each item is scored on a Likert scale from 1 to 5. A low score on this questionnaire reflects greater difficulty in processing sensory input. Scores are interpreted relative to the mean of the normative sample of children without disabilities (n=1037 [27]; Typical Performance (TP; within 1 standard deviation of the mean), Probable Difference (PD; below 1 standard deviation) or Definite Difference (DD; below 2 standard deviations). Reports of the analysis of the normative sample showed that age and gender differences were small and not meaningful for clinical application [27]. In the present study, the results were analysed using a one-sample t-test relative to the lower cut-off score for typical performance in the normative sample (specific mean values of the normative sample are not published in the manual). Participation was voluntary and approved by the board of directors of the AESWH. Members of the AESWH carried out the data collection. The researchers had no access to identifiable data.

RESULTS

Twenty-nine parents participated in the study providing information on the reactions to sensation of their children with WHS. The questionnaires from 12 parents were omitted from the analysis because their children fell outside the established

age range of the SSP (3 to 10 years). One questionnaire was omitted from the analysis due to missing data. The final analysis was carried out on the data provided by 16 parents representing 16 children with WHS aged 3 to 10 years (Table 1). A one-sample t-test was run to examine the difference between the mean scores of each SSP category of our sample of children with WHS and the cut-off score for typical performance in the normative sample. There were no outliers in the data of four of the SSP categories, as assessed by inspection of a box plot. There were two outliers in the Tactile Sensitivity and Movement Sensitivity categories, and in the Visual/Auditory Sensitivity category, there was one outlier. All outliers were low scores and we opted to include them in the analysis because verification showed they did not affect the result of the one-sample t-test (analysis with and without the outliers yielded the same result). Scores in five of the SSP categories were normally distributed, as assessed by Shapiro-Wilk's test ($p > .05$). Only the scores from the Tactile Sensitivity and Movement Sensitivity categories were not normally distributed. However, given that non-normality does not affect Type I error rate substantially and that the one-sample t-test can be considered fairly robust to deviations from normality [28], we opted to run the one-sample t-test regardless of this deviation. In all of the SSP categories, the group mean for children with WHS was below the Typical Performance cut-off score. The means of two categories (Underresponsive/Seeks Sensation, Low Energy/Weak) fell in the Definite Difference range and the rest of the category means fell in the Probable Difference range. However, the differences were statistically significant only for two categories: Underresponsive/Seeks Sensation and Low Energy/Weak (Table 2). These statistically significant differences were also found to be of significant magnitude with a large effect size ($> .8$) [29].

AGE (years)	Frequency	Percent
3	4	25.0
4	2	12.5
6	1	6.3
7	3	18.8
8	2	12.5
9	1	6.3
10	3	18.8
Total	16	100.0

SSP Category	Mean Score (SD) WHS (n=16)	Cut-off score T.P.	Difference	95% C.I.	t	p	d
Tactile Sensitivity	28.13 (5.82)	30	1.87	-1.23 to 4.98	-1.289	.217	-
Taste/Smell Sensitivity	12.94 (4.97)	15	2.06	-0.59 to 4.71	-1.659	.118	-
Movement Sensitivity	12.50 (3.52)	13	0.50	-1.38 to 2.38	-.568	.578	-
Under responsive. Seeks Sensation	21.06 (6.81)	27	5.94	2.31 to 9.56	-3.489	.003	.87
Auditory Filtering	19.94 (5.89)	23	3.06	-0.08 to 6.20	-2.079	.055	-
Low Energy/Weak	16.25 (7.44)	26	9.75	5.78 to 13.72	-5.240	.000	1.31
Visual/Auditory Sensitivity	17.63 (4.38)	19	1.37	-0.96 to 3.71	-1.256	.228	-

WHS: Wolf-Hirschhorn Syndrome; SD: Standard Deviation; TP: Typical Performance; CI: Confidence Interval; t = observed t value; p = p value (If $p > .05$, the difference is not statistically significant); d = Cohen's d calculated for values that were statistically significant

DISCUSSION

The present study offers a preliminary view on the presence of challenges in sensory integration and processing in children with WHS. The results point to the need to take into account sensory issues as a possible factor that contributes to the developmental and participation difficulties experienced by this population. Two categories of the SSP, Underresponsive/Seeks Sensation and Low Energy/Weak, were specifically identified as problematic in our sample of children with WHS. An atypical score in the Under-Responsive/Seeks Sensation section may be a manifestation of difficulties in registering or becoming aware of sensory information [21]. People with these kinds of difficulties often have a need to experience sensation at more intense levels than most individuals to become aware of sensory information [30,31]. Proxy questionnaires do not offer a complete understanding of sensory challenges, especially in the area of sensory perception, and direct assessment is needed to complement data collected from questionnaires such as the SSP [26,32]. However, observations of lack of awareness and seeking of sensory input may be an indication of difficulty perceiving sensation [19]. Given that adequate perception of sensory input is essential for adequate postural control and

learning new skills, consideration of challenges in sensory registration and perception should be a part of the assessment of children with WHS. Postural control and learning new skills are of particular concern in WHS [18]. For example, Sabbadini et al [18] reports that 91% of individuals were not self-sufficient in eating, dressing, washing or going to the bathroom.

The other category of the SSP found to be of concern in our sample was the Low Energy/Weak category. An atypical score in this section may be a manifestation of difficulties in processing sensory information from the muscles and joints (proprioceptive sensation) [19]. People with these types of difficulties often have weak muscles or problems of postural control [21]. Once again these issues are linked to the difficulties observed in clinical practice and reported in the literature. For example, Sabbadini et al. [18] and Battaglia et al. [33] report frequent difficulties in de-ambulation and functional control of the limbs. Understanding the relationship between sensory processing and integration and participation in daily occupations is complex [32]. Characterization of diagnostic groups is a first step in understanding this relationship but individual variations may be significant. An individualized assessment, carried out by an occupational therapist with advanced training in the assessment of sensory integration problems and its effects on participation in daily activities, is necessary [21,26,32]. Although we have focused on the two sections of the SSP that fell in the Definite Difference range and showed statistical difference with the cut off score for typical performance, results in the Probable Difference range also warrant our attention [21]. Scores in this range represent children in the lower 3-14% of the normative sample and it is probable that sensory processing difficulties interfere with performance in daily life [21].

LIMITATIONS AND FUTURE DIRECTIONS

The main limitation to this study lies in the fact that our sample was small. However, given that WHS is considered a rare condition, obtaining data from 16 children is of utmost importance. Additionally, we must consider that data was collected exclusively with a proxy questionnaire; no direct measures of sensory reactivity and perception were collected. This is definitely an important next step to better understand the sensory challenges faced by this population. Furthermore,

evidence supporting the use of occupational therapy in improving participation in children with sensory issues is growing [8,34,35]. However, none of the studies has included children with WHS, thus formally examining intervention in this population is needed.

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