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Sensory patterns of children with Williams syndrome

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Aim: Williams syndrome is a neurodevelopmental disorder caused by a deletion on chromosome 7. It is characterized by a range of medical problems in addition to the demonstration of maladaptive emotional and physical responses to environmental stimuli. Furthermore, sensory processing abnormalities are common in children with Williams syndrome. Therefore, this study aimed to report sensory processing difficulties in children with Williams syndrome in Turkey.

Methods: Twenty-three children with Williams syndrome (mean age 63.16±13.50 months; females n=13) and twenty-two typically developing children (mean age 67.66±13.23 months; females n=12) were included. Parents completed the Sensory Profile Questionnaire. Data were analysed using descriptive statistics and the Mann-Whitney U test.

Results: Children with Williams syndrome demonstrated sensory processing dysfunction in the sensory processing, modulation, behaviour and emotional response areas. Moreover, they have difficulties in low registration, sensation seeking, sensory sensitivity, sensation avoiding, sensory seeking, emotionally reactive low endurance/tone, oral sensitivity, inattention/distractibility, poor registration, sedentary behaviours, and fine motor/perceptual skills factors compared to their typically developing peers.

Conclusions: These findings, considered with similar published studies, confirm the prevalence and types of sensory processing abnormalities in Williams syndrome.

Keywords: CHILDREN; SENSORY PROCESSING; WILLIAMS SYNDROME; CHROMOSOME DISORDERS

INTRODUCTION

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a genetic deletion on the long arm of chromosome 7 (1). The prevalence is approximately 1 in 20,000 live births (1). WS is generally characterized by intellectual disability (usually mild) and delayed language and motor development (2). The WS personality profile involves high levels of sociability, an over-eagerness to interact with others, sensitivity, and tenseness (2). Children with WS also demonstrate maladaptive emotional and physical responses to environmental stimuli, including distractibility, ritualism, and indiscriminate social behaviour (3).

Sensory input from the environment and the body provides information to the brain. The brain organizes, integrates, synthesizes, and uses this information to understand experiences and organize appropriate responses. Information processing allows individuals to respond automatically, efficiently, and comfortably to the specific sensory inputs received (4). Sensory processing functions are crucial because they provide the basis for activities of daily living, learning, and motor development (5, 6). Furthermore, sensory processing helps children to show functional behaviour (7). Modulation of information needs to create an interchange along a continuum of habituation and sensitization (7).

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When a child has difficulty modulating between habituation and sensitization, they present with maladaptive behaviours such as hyperactivity, excessive lethargy, and inattentiveness (6). Like neurodevelopmental disorders, sensory processing abnormalities are common in children with WS. For instance, music-loving is a typical feature of individuals with WS (8); however, they show hyperresponses to some voices, such as hairdryers and vacuum cleaners, which seems paradoxical with this feature (9).

In recent years, there has been a growing body of evidence showing the presence of sensory processing difficulties in children with WS (10-14). *John and Mervis* (15) evaluated children's sensory processing skills with Short Sensory Profile and found that more than half of their sample was classified as definitely having overall sensory modulation issues. Other researchers have tried to show sensory processing abnormalities with Short Sensory Profile. They supported that overwhelmingly characterized behaviour and sensory processing issues can mainly be related to vestibular, auditory, gustatory, and proprioceptive functions (11, 16). Although recent studies showed that children with WS scored definite differences in sensory processing patterns, there is still a need for studies regarding sample size, using detailed sensory processing assessments, effects of comorbidities, effects of occupational therapy, and parental education. In light of the given literature, the current study aims to deeply evaluate the sensory patterns of children with WS and compare them with typically developing peers.

PATIENTS AND METHODS

Participants

Participants were enrolled in the Sensory Integration Unit Department of Occupational Therapy at Hacettepe University. Parents willing to participate in the study provided informed written consent on behalf of their children. The power sampling calculation indicated that the study's sample size of 20 participants would ensure 80% power and a 95% confidence interval for the study. Data from 23 children with WS between the ages of 3-10 years old were obtained, and over half of the sample were females. Each individual with WS had their diagnosis confirmed by a genetic test.

Measurements

The study and the control group assessments were performed by the first author and the second author, who were blinded to the group allocation. Each child and their parents were assessed individually in a quiet, distraction-free area in the clinic room. Demographic data were obtained from the interview.

TABLE 1. Demographic information of children with Williams syndrome (WS) and typical development (TD)

	Children with WS (n=23)	Children with TD (n=22)	z	p
Children's gender n (%)				
Female	13 (56.6%)	12 (54.6%)	-0.273	0.785
Male	10 (43.4%)	10 (45.4%)		
Children's chronological age in months (*M* ± *SD* (min–max))	63.16 ± 13.50 (40-101)	67.66 ± 13.23 (48-103)	-1.833	0.067
Mother's mean age, yr (*M* ± *SD*)	32 ± 4.37 (25-46)	32.13 ± 5.15 (23-42)	-0.066	0.948
Mother's education, n (%)				
Postgraduate	7 (30.4%)	5 (22.8%)		
Higher	1 (4.3%)	2 (9%)	-0.735	0.462
Secondary	15 (65.3%)	15 (68.2%)		
Spouse mean age, yr (*M* ± *SD*)	35.96 ± 4.42 (29-55)	36.47 ± 5.66 (27-49)	-0.338	0.736
Spouse education, n (%)				
Postgraduate	11 (47.9%)	10 (45.6%)		
Higher	4 (17.4%)	6 (27.2%)	-0.147	0.883
Secondary	8 (34.7%)	6 (27.2%)		
Family income, n (%)				
Low	4 (17.4%)	4 (18.1%)		
Average	10 (43.4%)	11 (50%)		
High	9 (39.2%)	7 (31.9%)	-0.609	0.543
Number of siblings, n (%)				
No sibling	4 (17.4%)	2 (9%)		
One sibling	9 (39.1%)	5 (23%)		
2-3 siblings	7 (30.4%)	13 (59%)	-1.566	0.117
More than 3 siblings	3 (13.1%)	2 (9%)		

The Sensory Profile Questionnaire assessed the sensory processing patterns of the children. This questionnaire consists of 125 items, in which parents report the frequency with which their child responds to items. The domains of the Sensory Profile Questionnaire are six sensory processing areas *(auditory, visual, vestibular, touch, multisensory and oral sensory),* five modulation areas *(endurance/tone, body position and movement, activity levels, emotional responses and visual input affecting emotional responses and activity level)* and three behaviour and emotional response areas *(emotional/social responses, behavioural outcomes of sensory processing, items indicating thresholds for response)*. This frequency is determined from a Likert scale from 1 to 5, where 1 always: when presented with the opportunity, the child responds in the manner described every time or 100% of

TABLE 2. Sensory Profile Questionnaire scores for children with Williams syndrome (WS) and typical development (TD)

Sensory Profile Questionnaire Quadrants, Factors and Subscales		Children with WS *M±SD* (min–max)	Children with TD *M±SD* (min–max)	z	p
Sensory Processing	Auditory Processing	23.61±5.34 (10-34)	32.54±5.07 (19-40)	-6.868	**0.001
	Visual Processing	29.16±6.03 (18-40)	38.35±4.47 (28-45)	-6.825	**0.001
	Vestibular Processing	40.44±5.63 (28-52)	48.62±6.33 (32-55)	-6.031	**0.001
	Touch Processing	69.40±6.94 (56-83)	80.73±9.31 (51-90)	-6.199	**0.001
	Multisensory Processing	23.42±5.22 (13-33)	30.20±3.46 (23-35)	-6.322	**0.001
	Oral Sensory Processing	39.90±11.25 (14-60)	48.60±8.86 (28-60)	-4.031	**0.001
Modulation	Sensory Processing Related to Endurance/Tone	30.27±7.77 (10-44)	41.47±3.19 (34-45)	-7.285	**0.001
	Modulation Related to Body Position and Movement	32.44±6.30 (18-46)	38.71±6.52 (21-50)	-4.544	**0.001
	Modulation of Movement Affecting Activity Level	21.37±4.62 (8-34)	29.16±4.61 (20-38)	-6.861	**0.001
	Modulation of Sensory Input Affecting Emotional Responses	11.42±3.00 (5-19)	16.49±2.95 (8-20)	-6.773	**0.001
	Modulation of Visual Input Affecting Emotional Responses and Activity Level	13.53±2.83 (5-20)	16.07±2.86 (10-20)	-4.272	**0.001
Behaviour and Emotional Responses	Emotional/Social Responses	58.55±9.74 (37-78)	67.69±9.63 (44-85)	-4.461	**0.001
	Behavioural Outcomes of Sensory Processing	17.40±4.89 (7-27)	24.00±4.80 (11-30)	-5.874	**0.001
	Items Indicating Thresholds for Response	11.01±2.15 (5-15)	13.16±2.36 (6-15)	-4.903	**0.001
Quadrants	Low Registration	50.29±10.48 (19-69)	65.86±5.82 (54-75)	-7.283	**0.001
	Sensation Seeking	87.40±12.62 (59-114)	125.13±26.68 (65-169)	-7.006	**0.001
	Sensory Sensitivity	68.07±11.09 (43-93)	85.96±10.36 (59-99)	-6.648	**0.001
	Sensation Avoiding	102.29±14.16 (65-131)	124.00±13.89 (91-146)	-6.466	**0.001
Factors	Sensory Seeking	55.27±10.51 (31-79)	68.15±10.56 (36-85)	-5.582	**0.001
	Emotionally Reactive	51.11±11.09 (28-74)	62.50±10.31 (37-80)	-4.869	**0.001
	Low Endurance/Tone	30.42±8.91 (10-50)	41.47±3.19 (34-45)	-6.719	**0.001
	Oral Sensitivity	29.50±8.69 (11-45)	35.15±8.08 (13-45)	-3.314	**0.001
	Inattention/Distractibility	20.31±4.83 (8-31)	28.88±4.17 (20-35)	-7.239	**0.001
	Poor Registration	32.20±3.20 (17-40)	35.50±4.24 (21-40)	-4.621	**0.001
	Sensory Sensitivity	12.31±4.08 (6-20)	17.00±2.50 (8-20)	-5.884	**0.001
	Sedentary Behaviours	13.27±3.56 (4-20)	17.07±2.88 (10-20)	-5.273	**0.001
	Fine Motor/Perceptual Skills	7.53±2.33 (3-13)	12.45±2.18 (8-15)	-7.718	**0.001

**p<0.01; z: Mann-Whitney U test

the time; 2: frequently, or at least 75% of the time; 3: occasionally, or 50% of the time; 4: seldom, or 25% of the time; and 5: never: when presented with the opportunity, the child never responds in this fashion, or 0% of the time (17). The cross-cultural adaptation of the questionnaire was carried out, and Cronbach's alpha ranged from 0.63 to 0.97 with excellent test/retest reliability (18).

Statistical Analysis

Data were analysed using IBM Statistical Package for the Social Sciences (SPSS) version 23.0 software. The variables were investigated using visual (plots/histograms) and analytical methods (Kolmogorov-Smirnov test) to determine whether they were normally distributed. Descriptive statistics were expressed as the mean ± standard deviation or median (min–max) according to the assumption of a normal distribution. The Mann-Whitney U test was used to compare the measurements between groups. All results were given as the mean ± SD, median (M), range and p values <0.05 considered to be significant.

RESULTS

The demographic characteristics of the children are shown in Table 1. There were no statistically significant differences between the groups in terms of demographic information (p>0.05). The Sensory Profile Questionnaire scores between the groups were compared by examining the performance of each group on each factor. Table 2 presents each group's Sensory Profile Questionnaire scores. The table indicates the mean scores, standard deviations, minimum and maximum

scores received according to groups. There was a statistically significant difference between the groups in the subscales ($p<0.05$).

DISCUSSION

The results of this study confirmed our hypothesis that the scores of the Sensory Profile Questionnaire of children with WS were significantly different from those of children with typical development. Most children with WS demonstrate different sensory patterns in sensory modulation. Moreover, they have difficulties in sensory seeking, emotional reactivity, low endurance/tone, oral sensitivity, inattention/distractibility, sensory sensitivity and fine motor/perceptual factors.

Children with WS showed sensory seeking, which is related to hyperactivity and inattention behaviour during occupational performance. This result is consistent with the literature and supported by several authors (19). Our results showed that children with WS are more emotionally reactive than children with typical development, which causes poor coping and variability in emotional responses during occupational performance. This result is supported by the study of *Klein-Tasman and Mervis* (2), which confirmed that the WS personality profile was characterized by a combination of high sensitivity to criticism and high anxiety. According to our results, children with WS showed low endurance and muscle tone more than those with typical development in the SP questionnaire, which can cause hypotonia. The literature is consistent with our results, and it is clear that children with WS have lower postural stability and core strength than their typically developing peers, and their antigravity muscle strength is lower than that of their typical peers (15, 20, 21). It is assumed that low endurance and tone, a characteristic feature of the WS phenotype (2, 22), can be related to unusual sensory responses, especially in the vestibular sense, which we found among children with WS.

According to our results, children with WS showed oral sensitivity. Picky eating is often characterized by strong food preferences and rejection of familiar or new foods (23), and children with low tactile awareness tend to show picky eating habits. The literature supports that children with WS show decreased/increased food repertoire (3). Moreover, picky eating can be related to tactile sense. Our results showed that the tactile sense and awareness of children with WS are lower than those of their typically developing peers. Accordingly, increased/decreased oral sensitivity seen among children with WS may be related to tactile awareness of children with WS.

Our results supported that the children with WS showed increased sensory sensitivity. It is well supported that children with WS demonstrate hypersensitivity to sound and have poor auditory filtering (1, 15, 24, 25).

Additionally, *John and Mervis* (15) documented that parents reported that their child with WS demonstrated auditory aversions. Moreover, it is supported that children with WS had difficulties with visual stimuli and visual perception (25, 26). Our results confirm that children with WS have lower capacities to adapt tactile, vestibular, auditory, visual and multisensory processing than their typical peers. It is thought that WS children's sensory sensitivity is related to their poor tactile, vestibular, auditory, visual and multisensory processing skills.

According to our results, the fine motor skills and perceptual skills of children with WS are lower. It was supported that children with WS had difficulties in fine motor development because of visual motor deficits (27, 28). Our results are consistent with the literature by showing that children with WS have lower visual processing skills. However, our results also show different relationships, such as decreased proprioceptive and body awareness, which can cause a delay in fine motor skills. Therefore, a better understanding of the complex relationship between fine motor skills and sensory processing skills in children with WS is an important area for future research.

Our results confirmed that children with WS show increased inattention and distractibility behaviour during occupational performance. Play is the primary occupation of children. They can build their learning, imitation capacities, gross/fine motor and language skills during play. Inattention and distractibility can cause decreased play skills and negatively affect learning, imitation and language skills. Moreover, sensory modulation problems can affect play skills (29). There is no study investigating the sensory modulation and play skills of children with WS in the literature. However, our results confirm that increased inattention and distractibility can be related to our finding of decreased sensory modulation skills in children with WS. This result may be related to the decreased learning and play skills of children with WS.

The sensory processing findings noted in this study reflect a pattern of dysfunctional sensory modulation; that is, children with WS demonstrate difficulty with filtering and changing to sensory stimuli to develop an adaptive response during daily living. Sensory modulation has been defined as the capacity to regulate and organize the degree, intensity, and nature of responses to sensory input in a graded and adaptive manner (29, 30). In turn, sensory modulation allows a person to achieve and maintain an optimal range of performance and to adapt to daily life challenges.

CONCLUSION

Sensory modulation is essential for higher-level learning, adaptive behaviour, and social functioning. The results of

the present study indicated that most children with WS demonstrate abnormal sensory patterns, mainly in the sensory modulation area. More research is needed to examine the nature and impact of sensory modulation abnormalities on the behavioural phenotype associated with WS. In particular, while the present results indicate that sensory modulation difficulties are a key component of the WS phenotype, the developmental progression of sensory modulation difficulties is unknown. Sensory modulation can be affected by sociodemographic variables such as age and gender in neurodevelopmental disorders such as autism spectrum disorders. Furthermore, additional research about the play skills of children with WS is needed to understand the effects of sensory processing dysfunction in children with WS.

REFERENCES

1. Martens MA, Wilson SJ, Reutens DC. Research review: Williams syndrome: a critical review of the cognitive, behavioral, and neuroanatomical phenotype. J Child Psychol Psychiatry. 2008;49:576–608.
2. Klein-Tasman BP, Mervis CB. Distinctive personality characteristics of 8-, 9-, and 10-year-olds with williams syndrome. Dev Neuropsychol. 2003;23:269–290.
3. Mervis CB, Klein-Tasman BP. Williams syndrome: cognition, personality, and adaptive behavior. Dev Disabil Res Rev. 2000;6:148-158.
4. Ayres AJ. Types of sensory integrative dysfunction among disabled learners. Am J Occup Ther. 1972;26:13–18.
5. Celik HI, Elbasan B, Gucuyener K, Kayıhan H, Huri M. Investigation of the relationship between sensory processing and motor development in preterm infants. Am J Occup Ther. 2018;72:7201195020p1-7201195020p7.
6. Dunn W. The impact of sensory processing abilities on the daily lives of young children and their families: a conceptual model. Infants Young Child. 1997;9:23-35.
7. Dunn W. Sensory Profile: Examiner's Manual. The Psychological Corporation, San Antonio; 1999.
8. Carrasco X, Castillo S, Aravena T, Rothhammer P, Aboitiz F. Williams syndrome: pediatric, neurologic, and cognitive development. Pediatr Neurol. 2005;32:166-172.
9. Pober BR. Williams–Beuren syndrome. N Engl J Med. 2010;362:239-252.
10. Powell B, Van Herwegen J. Sensory processing in Williams syndrome: individual differences and changes over time. J Autism Dev Disord. 2022;52:3129-3141.
11. Glod M, Riby DM, Rodgers J. Relationships between sensory processing, repetitive behaviors, anxiety, and intolerance of uncertainty in autism spectrum disorder and williams syndrome. Autism Res. 2019;12:759-765.
12. Glod M, Riby DM, Rodgers J. Sensory processing in Williams syndrome: a narrative review. Rev J Autism Dev Disord. 2020;7:32-45.
13. Glod M, Riby DM, Rodgers J. Sensory processing profiles and autistic symptoms as predictive factors in autism spectrum disorder and williams syndrome. J Intellect Disabil Res. 2020;64:657-665.
14. Riby DM, Janes E, Rodgers J. Brief report: exploring the relationship between sensory processing and repetitive behaviours in williams syndrome. J Autism Dev Disord. 2013;43:478-482.
15. John AE, Mervis CB. Sensory modulation impairments in children with williams syndrome. Am J Med Genet C: Semin Med Genet. 2010;154:266-276.
16. Janes E, Riby DM, Rodgers J. Exploring the prevalence and phenomenology of repetitive behaviours and abnormal sensory processing in children with williams syndrome. J Intellect Disabil Res. 2014;58:746-757.
17. Ermer J, Dunn W. The sensory profile: a discriminant analysis of children with and without disabilities. Am J Occup Ther. 1998;52:283-290.
18. Kayıhan H, Akel BS, Salar S, et al. Development of a turkish version of the sensory profile: translation, cross-cultural adaptation, and psychometric validation. Percept Mot Skills. 2015;120:971-986.
19. Braga AC, Carreiro LRR, Tafla TL, et al. Cognitive and behavioral profile of williams syndrome toddlers. CoDAS. 2018;30:e20170188.
20. Barozzi S, Soi D, Gagliardi C, et al. Balance function in patients with williams syndrome. Gait Posture. 2013;38:221-225.
21. Mazzocco MM, Ross JL, eds. Neurogenetic developmental disorders: Variation of manifestation in childhood. MIT Press; 2007.
22. Lincoln AJ, Searcy YM, Jones W, Lord C. Social interaction behaviors discriminate young children with autism and williams syndrome. J Am Acad Child Adolesc Psychiatry. 2007;46:323-331.
23. Taylor CM, Wernimont SM, Northstone K, Emmett PM. Picky/fussy eating in children: review of definitions, assessment, prevalence and dietary intakes. Appetite. 2015;95:349-359.
24. Zarchi O, Attias J, Gothelf D. Auditory and visual processing in williams syndrome. Isr J Psychiatry Relat. 2010;47:35-41.
25. Marler JA, Elfenbein JL, Ryals BM, Urban Z, Netzloff ML. Sensorineural hearing loss in children and adults with williams syndrome. Am J Med Genet A. 2005;138:318-327.
26. Farran EK, Jarrold C. Visuospatial cognition in williams syndrome: reviewing and accounting for the strengths and weaknesses in performance. Dev Neuropsychol. 2003;23:173-200.
27. Heiz J, Barisnikov K. Visual–motor integration, visual perception and motor coordination in a population with williams syndrome and in typically developing children. J Intellect Disabil Res. 2016;60:945-955.
28. Gagliardi C, Martelli S, Burt MD, Borgatti R. Evolution of neurologic features in Williams syndrome. Pediatr Neurol. 2007;36:301-306.
29. Ismael N, Lawson LM, Hartwell J. Relationship between sensory processing and participation in daily occupations for children with autism spectrum disorder: a systematic review of studies that used Dunn's sensory processing framework. Am J Occup Ther. 2018;72:7203205030p7203205031-7203205030p7203205039.
30. Kennedy-Behr A, Rodger S, Mickan S. A comparison of the play skills of preschool children with and without developmental coordination disorder. OTJR Occup Particip Health. 2013;33:198-208.

SAŽETAK

Senzorni obrasci djece s Williamsovim sindromom

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Cilj: Williamsov sindrom je neurorazvojni poremećaj uzrokovan delecijom na kromosomu 7. Karakterizira ga niz medicinskih problema uz demonstraciju neprilagodljivih emocionalnih i fizičkih odgovora na podražaje iz okoline. Nadalje, poremećaj senzorne integracije česte su kod djece s Williamsovim sindromom. Stoga je ova studija imala za cilj izvijestiti o poteškoćama senzorne integracije kod djece s Williamsovim sindromom u Turskoj.

Metode: Uključeno je dvadeset troje djece s Williamsovim sindromom (prosječne dobi 63,16±13,50 mjeseci; ženskog spola n=13) i dvadeset dvoje djece u tipičnom razvoju (prosječne dobi 67,66±13,23 mjeseci; ženskog spola n=12). Roditelji su ispunjavali upitnik senzornog profila (Sensory Profile Questionnaire). Podaci su analizirani pomoću deskriptivne statistike i Mann-Whitney U testa.

Rezultati: Djeca s Williamsovim sindromom pokazala su poremećaj senzorne integracije u područjima senzorne obrade, modulacije, ponašanja i emocionalnog odgovora. Štoviše, imaju poteškoće u slabom registriranju, traženju osjeta, senzornoj osjetljivosti, izbjegavanju osjeta, senzornom traženju, emocionalno reaktivnoj niskoj izdržljivosti/tonusu, oralnoj osjetljivosti, nepažnji/distraktibilnosti, slabom registriranju, sjedilačkom ponašanju i poteškoćama fine motorike/perceptivnih vještina u usporedbi s njihovi vršnjaci u tipičnom razvoju.

Zaključci: Ovi nalazi, uzeti u obzir sa sličnim objavljenim studijama, potvrđuju prevalenciju i vrste poremećaja senzorne integracije u Williamsovom sindromu.

Ključne riječi: DJECA; SENZORNA INTEGRACIJA; WILLIAMSOV SINDROM; KROMOSOMOPATIJE