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Cognitive Abilities in Patients with Williams Syndrome

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Objective: Patients with Williams syndrome (WS) show intellectual disabilities and specific cognitive profiles have been suggested. However, we hypothesized that these features might not be common to all patients with WS. We performed this study to test our hypothesis.

Subjects and Methods: The subjects were 22 WS patients 6-30 years of age (mean ± standard deviation, 13.14 ± 7.00 years). Each subject was administered the Wechsler Intelligence Scale (Wechsler test) and/or the Kaufman Assessment Battery for Children (K-ABC). A cluster analysis was conducted to identify performance profiles based on the K-ABC subtest scores by Ward's hierarchical agglomerative method. As we hypothesized, 2 clusters were recognized in the preliminary results. Finally, both clusters were statistically analyzed.

Results: According to patient age, several types of examinations were performed; 19 underwent the Wechsler test, 2 the Tanaka Binet test, and 21 the K-ABC. The subtest profile revealed that the subjects exhibited higher comprehension scores, while the mean score on block design was lowest. Two clusters, designated groups A and B, were constructed by Ward's hierarchical agglomerative method from the 21 subjects administered the K-ABC. There was a statistically significant difference between the 2 clusters on all subtests of sequential processing and on 2 subtests of simultaneous processing (gestalt closure and triangle), with $p<0.05$ for hand movement and number recall, $p<0.01$ for word order and gestalt closure, and $p<0.0001$ for triangle. In contrast, these two groups showed no significant differences in matrix analogies or spatial memory scores.

Conclusion: On K-ABC, WS patients showed a wider range of the results. From this experience, we considered the triangle task potentially be suitable for detecting the degree of visuo-spatial disability in individuals with WS. Although weakness on the spatial ability subtest was a common finding in WS patients, the spatial memory test apparently had the sensitivity to detect this core weakness in patients with WS. In conclusion, we confirmed our hypothesis that patients with WS show variability in cognitive profiles.

Key Words: Williams syndrome, cognitive profile, Wechsler Intelligence Scale, Kaufman Assessment Battery for Children

Introduction

Williams syndrome (WS) is a genetic disorder, affecting multiple organ systems, caused by a hemizygous microdeletion in the long arm of chromosome 7 (7q11.23)[1)2)]. WS is characterized by elastin arteriopathy, dysmorphic features, and mental deficits or learning difficulties. Patients with WS suffer from striking neurocognitive and social-behavioral abnormalities, making it difficult for some to cope with societal expectations[3)4)].

Much attention has been paid to the intellectual abilities of patients with WS who show mild to moderate mental deficits[5)~9)]. Several studies have indicated that language ability is relatively preserved[5)~7)10)~12)] and another suggested minor impairments in both linguistic and non-linguistic abilities in WS patients[9)]. Studies on the cognitive characteristics of WS patients have provided variable and conflicting results. Although it has been recognized that patients with WS demonstrate severe visuo-spatial deficits[5)10)13)] and impairments of visuo-spatial constructive cognition, relative strengths are seen in short-term verbal memory and language[5)11)12)14)15)]. Such specific cognitive profiles have been discov-

ered and confirmed in the majority of WS patients. However, we hypothesized that they might not be present in all patients with WS. Accordingly, this study was designed to test our hypothesis by examining whether patients with WS show core cognitive ability characteristics and whether each of these deficits is common to the vast majority of individuals with WS.

Subjects and Methods

The subjects were 22 patients with a diagnosis of WS based on clinical and genetic findings. The subjects ranged in age from 6 to 30 years (mean age ± standard deviation (SD), 13.14 ± 7.00 years). The Wechsler preschool and primary scale of intelligence (WPPSI), Wechsler Adult Intelligence Scale-Revised (WAIS-R) and Wechsler Intelligence Scale for Children III (WISC III) tests were conducted in one, 3 and 15 subjects, respectively. In two cases, the Tanaka-Binet Intelligence scale (Japanese version of the Stanford-Binet Intelligence scale) was conducted. Results for verbal intelligence quotient (VIQ) and performance intelligence quotient (PIQ), as well as subtest standard scores, were compared.

In 21 subjects (one of whom, unfortunately, did not undergo intelligence scale testing), the Kaufman Assessment Battery for Children (K-ABC)[16)17)] was performed. The K-ABC has no age-equivalent scales for patients less than 13 years of age. Therefore, subjects up to age 13 were compared to those up to 12 years and 11 months of age. Both tests were administered by clinical psychologists. Though the K-ABC includes 14 subtests, in this study, we conducted 7 subtests, 3 sequential processing subtests (hand movement, number recall, word order) and 4 simultaneous processing subtests (gestalt closure, triangle, matrix analogies, spatial memory). Subjects administered the K-ABC were entered into a cluster analysis based on all included subtest standard scores of the K-ABC mental processing domains. Ward's hierarchical agglomerative method was used as the grouping procedure. Two clusters were recognized by the analysis, and several characteristics of patients in the two groups were also compared.

Table 1 Age and Full IQ (FIQ) Distribution of Subjects with Williams syndrome (n = 19)

Age (years)	Group	FIQ score				Total
		-40	41-50	51-70	71-	
6-9		3 (42.9)	2 (28.6)	2 (28.6)		7 (36.8)
10-12		0 (0)	3 (50)	2 (33.3)	1 (16.7)	6 (31.6)
13-15		1 (100)	0 (0)	0 (0)		1 (5.3)
16-		1 (20)	2 (40)	2 (40)		5 (26.3)
Total (%)		5 (26.3)	7 (36.8)	6 (31.6)	1 (5.3)	19 (100)

Results

1. Wechsler Intelligence Test (Table 1-3, Fig. 1)

Table 1 and Fig. 1 present the age and full intelli-

Table 2 Verbal IQ (VIQ), Performance IQ (PIQ) and Full (IQ) in Williams syndrome (n = 19)

IQ	Mean (SD)	Range	
VIQ	59.37 (10.71)	47-82	Student t = 5.731
PIQ	49.05 (8.34)	40-68	p < 0.0001
FIQ	49.58 (9.08)	40-73	

Table 3 Wechsler intelligence subtests in Williams syndrome (n = 19)

		Mean (SD)	Range	Difference between means (Simultaneous 95% confidence limits)	
VIQ	Information	2.84 (1.68)	1-7	*2.52 (0.80-4.25)	
	Similarities	3.79 (3.12)	1-10	*1.89 (0.16-3.62)	F = 8.02
	Arithmetic	2.21 (1.69)	1-7	*3.16 (1.43-4.88)	p < 0.0001
	Vocabulary	3.47 (2.06)	1-7	*1.89 (0.16-3.62)	
	Comprehension	5.37 (2.48)	1-8	*1.95 (0.22-3.67)	
PIQ	Picture completion	3.42 (2.29)	1-8	*3.47 (1.75-5.20)	
	Block design	1.89 (1.79)	1-8		

Comparisons significant at 0.05 level are indicated by*. (Tukey's studentized range test for subtests)
Subject's performance was substantially better in the comprehension subtest. Mean scales were 5.37 (SD = 2.48) and 3.79 (SD = 3.12) for the comprehension and similarities subtests respectively.
The mean scaled for block design was 1.89 (SD = 1.79).

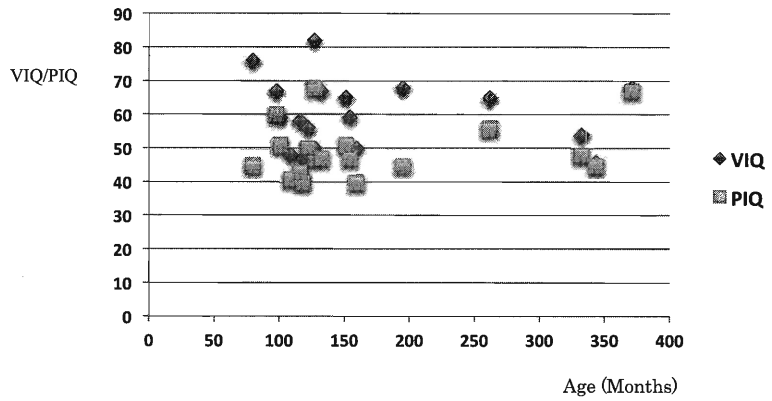

Fig. 1 Age and IQ Distributions of Subjects with Williams syndrome (n = 19)

Table 4 Mean K-ABC standard scores in 21 Williams syndrome subjects

	Mean (SD)	Range	
K-ABC scale			
Sequential processing	70.52 (17.15)	45-105	Student t = 3.95331
Simultaneous processing	57.95 (12.20)	43-91	p = 0.0008
Achievement	65.29 (11.28)	49-97	
Achievement subtests			
Riddles	66.95 (6.61)	58-85	
Arithmetic	64.19 (15.37)	45-90	
Reading/decoding	62.95 (14.38)	45-112	
Reading/understanding	76.14 (14.13)	56-114	

Table 5 Mean K-ABC scaled scores and standard deviation in 21 Williams syndrome subjects

	Mean (SD)	Range	Difference between means (Simultaneous 95% confidence intervals)	
Sequential processing subtest				
Hand movement	5.24 (3.51)	1-12	*2.90 (0.94-4.87)	
Number recall	4.71 (3.70)	1-16	*2.43 (0.46-4.39)	
Word order	6.14 (3.68)	1-14	*2.38 (0.42-4.34)	
Simultaneous processing subtest			*3.81 (1.85-5.77)	F = 8.76
Gestalt closure	2.33 (1.43)	1-5	*2.24 (0.28-4.20)	p < 0.0001
Triangle	3.90 (2.68)	1-10	*2.81 (0.85-4.77)	
Matrix analogies	3.33 (1.74)	1-7	*3.33 (1.37-5.30)	
Spatial memory	2.81 (1.69)	1-7		

Comparisons significant at 0.05 level are indicated by*. (Tukey's studentized range test for subtests)

gence quotient (FIQ) scores of the subjects. FIQ scores suggested that 18 subjects (95%) were mentally deficient and 12 (63%) had FIQ < 50. VIQ scores were significantly higher than PIQ scores as shown in Table 2. There was a significant difference between subtest scores ($F = 8.02$; $p < 0.0001$) by two-way analysis of variance (independent variables: subtest and case) and mean subtest results are displayed in Table 3. Tukey's HSD test showed the comprehension subtest mean score to be highest (5.37) with significant differences ($F = 8.02$, $p < 0.0001$) from all other subtests. The mean scaled score for similarities (3.79) was the second highest. Among all subtests, the mean score for block design was lowest (1.89).

2. K-ABC scores (Table 4, 5)

The mean K-ABC standard scores of 21 subjects are shown in Table 4. Sequential processing and achievement were scaled higher than simultaneous processing. The mean scale was 70.52 (SD = 17.15)

for sequential processing and 57.95 (SD = 12.20) for simultaneous processing, and the difference was statistically significant (p = 0.0008). Among achievement subtests, the subjects had relatively high scores on the reading/understanding subtests.

The mean K-ABC scaled scores of processing subtests and standard deviations are shown in Table 5. With simultaneous processing, mean scaled scores showed a significant difference (F = 8.76, p< 0.0001) between word order and the other subtests of simultaneous processing (gestalt closure, triangle, matrix analogies and spatial memory).

Table 6 Demographics and test scores of the two groups

Group	A group (n = 9)	B group (n = 12)	p
Sex M/F (n)	3/6	3/9	0.497
Age (years)	13.2 ± 7.7	12.2 ± 5.8	0.583
Wechsler test			
VIQ	65.5 ± 9.5	53.8 ± 9.2	0.017*
PIQ	53.4 ± 10.1	44.8 ± 3.4	0.027*
FIQ	54.9 ± 10.0	42.7 ± 5.0	0.017*
Tanaka-Binet			
IQ	34, 35		

*: p<0.05
Pearson's chi-square test for sex.
There were no significant differences in sex or age between the two groups.
Comparisons significant at the 0.05 level by paired t-test are indicated.
There are significant differences in VIQ, PIQ and FIQ between the two groups.
Three B group cases were excluded from this analysis.
In two cases, the Tanaka-Binet Intelligence Scale (Japanese version of the Stanford-Binet Intelligence Scale) was conducted, and the FIQ scores were 34 and 35. In one B group case, Intelligence Scale testing was not conducted.

3. Cluster analysis by Ward's hierarchical agglomerative method (Table 6-8, Fig. 2)

Based on all scaled processing scores of K-ABC subtests, subjects were classified into 2 clusters, designated groups A (n = 9) and B (n = 12). One of the clusters (group A) showed significantly higher scores on the "sequential processing" than on the "simultaneous processing" subtest. The cluster which was good at "sequential processing" tasks was group A.

Table 6 presents a comparison of gender, age, and IQ scores in these 2 clusters. Although there was no significant difference in gender or age, significant differences were identified in VIQ, PIQ and FIQ between the 2 groups.

Table 7 presents comparisons of K-ABC processing subtests between the 2 groups. In group B, low scores were identified in subtests of "sequential processing"; i.e., a score of 3.42 (SD = 1.88) for hand movement, 3.00 (SD = 1.54) for number recall and 4.08 (SD = 2.11) for word order. In group A, scores were higher, i.e., 7.67 (SD = 3.77) for hand movement and 8.89 (SD = 3.59) for word order. From these results, significant scaled score differences were confirmed between the 2 groups in all "sequential processing" tasks. In both groups, all subjects had low scores for "simultaneous processing" tasks.

There was no significant difference between the 2 groups in "simultaneous processing" subtests regarding matrix analogies and spatial memory, despite a significant difference between the 2 groups

Table 7 Comparison of K-ABC processing subtests between two groups (n = 21)

	A group (n = 9) Mean (SD)	B group (n = 12) Mean (SD)	Difference Mean (SD)
Sequential processing subtest (scaled score)			
Hand movement	7.67 (3.77)	3.42 (1.88)	4.25 (2.84)*
Number recall	7.00 (4.56)	3.00 (1.54)	4.00 (3.18)*
Word order	8.89 (3.59)	4.08 (2.11)	4.81 (2.83)**
Simultaneous processing subtest (scaled score)			
Gestalt closure	3.33 (1.41)	1.58 (0.90)	1.75 (1.15)**
Triangles	6.44 (1.88)	2.00 (1.13)	4.44 (1.49)****
Matrix analogies	3.44 (2.07)	3.25 (1.54)	0.19 (1.78)
Spatial memory	3.56 (1.81)	2.25 (1.42)	1.30 (1.60)

*: p<0.05, **: p<0.01, ***: p<0.001, ****: p<0.0001

Table 8 Comparison of mean K-ABC standard scores and achievement subtests between two groups

	A group (n = 9) Mean (SD)	B group (n = 12) Mean (SD)	Difference Mean (SD)
K-ABC scales (standardized score)			
Sequential processing	86.11 (12.03)	58.83 (9.03)	27.28 (10.40)****
Simultaneous processing	64.56 (13.48)	53.00 (8.73)	11.56 (10.98)*
Achievement	73.78 (10.11)	58.97 (7.33)	14.86 (8.61)***
Achievement subtests (standardized score)			
Riddles	69.89 (8.31)	64.75 (4.09)	5.14 (6.23)
Arithmetic	77.00 (7.23)	54.58 (12.52)	22.42 (10.62)****
Reading/decoding	71.33 (16.10)	56.67 (9.31)	14.67 (12.62)*
Reading/understanding	86.22 (14.18)	68.58 (8.45)	17.64 (11.26)**

*: $p<0.05$, **: $p<0.01$, ***: $p<0.001$, ****: $p<0.0001$

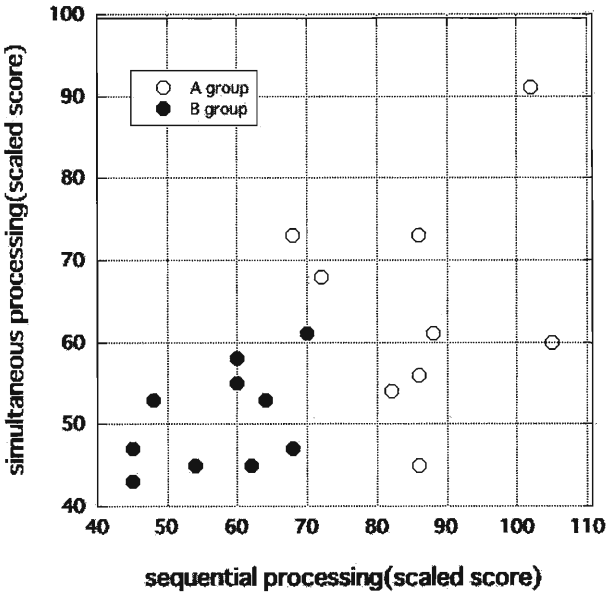

Fig. 2 Sequential processing/simultaneous processing of two groups

for gestalt closure ($p < 0.001$) and triangle ($p < 0.0001$). These WS patients especially showed weak cognitive ability when holistic and integrative problem-solving approaches were used. Spatial ability weakness may be a relatively common characteristic seen in most patients with WS, and spatial memory tasks may have the sensitivity to detect this core weakness.

Table 8 presents comparisons of mean K-ABC standard scores and achievement subtests between the 2 groups. Group A had a higher "sequential processing" score, i.e. 86.11 (SD = 2.03) as compared to only 58.83 (SD = 9.03) in group B. Regarding the results of subtests, group A showed significantly higher scores ($p<0.0001$) for the achievement tasks, particularly arithmetic. In both group A and group B, most subjects had good scores on the reading/understanding subtest, while scores were lower on the reading/decoding subtest. During the examination for the reading/decoding subtest, subjects are required to express the meanings of a sentence by employing gestures. These results suggested that WS patients have difficulty in reading each word, but that they are able to understand the meaning of the sentence.

Discussion

Most of our WS patients showed mild to moderate mental impairments, which are consistent with the results of previous studies[5)~7)12)]. We confirmed VIQ to be higher than PIQ in the patients with WS, as have many other studies[5)12)]. Other studies obtained low scores in the information, arithmetic and comprehension subtests, while scores for the similarities and vocabulary subtests were high[5)12)16)18)]. However, relatively high scores on comprehension subtests have rarely been reported in patients with WS. On the comprehension subtest, subjects are asked about a social situation or common concepts. This subtest includes questions such as what you should do if you notice a purse is left on the floor in a shop, why someone should turn off the light when nobody is in a room, and so on. The comprehension subtest may evaluate certain aspects of socialization ability. This might explain why comprehension results vary among patients and how some are able to understand daily life situations and social rules

while others cannot.

The score for the block design subtest was significantly lower than those for the similarities and comprehension subtests. On the block design subtest, subjects are required to replicate objects into a block design model. Low scores were scaled on tasks involving the integration and construction of visuo-spatial information in WS patients[12)13)]. Ferrero et al[19)] reported a patient with a mild WS physical phenotype and normal IQ. On the WPPSI test, this patient displayed strengths on all verbal subtests except for arithmetic and showed weaknesses on object the assembly, geometric designs and block design subtests. Block design involves the task of copying small geometric designs with cubes, and patients with WS may not be able to determine the block positions. This study confirmed spatial orientation to be impaired in WS patients. Low scores on tasks for PIQ subtests may be derived from impairments of visuo-spatial cognition. These results have also been confirmed in previous studies of WS patients showing language to be relatively well-preserved while there is a visuo-spatial cognition weakness.

A comparative strength in "sequential processing" was observed in this study. All subjects in both groups, which we classified according to the results of the K-ABC test, had higher scores on the hand movement, number recall, and word order subtests than on those of "simultaneous processing". In particular, the word order scores were highest. The subjects responded well to word sequences, as compared to sequences of numbers or hand movements. The word order task requires short-term memory of sequential words, and subjects are required to remember and transpose a corresponding picture. During this task, WS patients could remember the word linguistically, and may have been able to transpose the word meaning to the corresponding picture. The subjects in group A had sufficient ability to solve problems in temporal order. They could identify the picture corresponding to the words. We found that the group A subjects may have adequate visuo-feature cognition, allowing them to respond well on the word order task, as well as good verbal processing skills, as compared to those in group B. As we hypothesized, not all WS patients have good short-term memory.

A strong relationship was observed between the results of the Wechsler performance scale and the K-ABC simultaneous scale[20)], and impairments of visuo-spatial cognition in WS patients were demonstrated by the latter battery. Block design is one of the subtests of the Wechsler test, and triangle is one of the K-ABC subtests. In this study, all subjects had low scores on the block design subtest but group B subjects had the lowest scores. Use of the block design and triangle subtests allows similar cognitive skills to be measured in WS patients.

The triangle subtest requires planning ability and depends on various forms of psychological processing; however, the block design task requires more planning skill, which would account for the large difference between the 2 groups (A&B) on the block design subtest. Because WS patients showed a wider range of results for the triangle task subtest, it would potentially be suitable for detecting the degree of visuo-spatial disability in individuals with WS. Subjects of both groups had low spatial memory scores, reflecting their core cognitive dysfunction.

Using the cluster analysis method, we revealed the common characteristics of visuo-spatial deficiency in patients with WS. Good short-term memory was not common to all WS patients. Rather, a portion of the subjects, which we classified as group A, showed higher scores for short-term memory.

Conclusions

This study demonstrated VIQ to be superior to PIQ in WS patients, results consistent with those of the previous studies. However, the high comprehension scores observed in this study were striking. Using cluster analysis of K-ABC results, the subjects were classified into 2 groups, one of which showed significantly better scores on "sequential processing" rather than "simultaneous processing". This suggested that some WS patients have good short-term auditory memory. The common difficulty for most subjects was reflected in their low spatial memory scores. We consider the spatial

memory subtest of the K-ABC to be sufficiently sensitive to detect the core cognitive deficits in WS patients while the triangle subtest would be more informative regarding their visuo-spatial disability.

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ウィリアムズ症候群の認知の特徴

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〔はじめに〕Williams 症候群（WS）は 7 番染色体長腕の半接合体（7q11.23）の微小欠失によっておこる遺伝子疾患である．エラスチン動脈症，特徴的顔貌，知的障害あるいは学習障害が特徴的である．我々はこれまでの経験から，WS 患者の認知機能の障害は一律ではなく，いくつかのタイプに分けられるのではないかと考えていた．そこで，我々の仮説を検証するために本研究を行った．

〔目的〕WS の認知機能の核となる特徴と，そのうち多くの WS に共通して存在する特徴を明らかにする．

〔対象と方法〕本疾患 22 例，6〜30 歳（平均 13.14±7.00 歳）．19 人に Wechsler 系知能検査（Wechsler），2 人に田中ビネーを施行し，さらに 21 例は日本版 Kaufman 心理・教育アセスメントバッテリー（K-ABC）を施行した．K-ABC の下位項目の点数を基にクラスター解析（Ward's hierarchical agglomerative 法）を行ったところ我々の仮説どおり 2 群に分けられたので 2 群の検討をした．

〔結果〕知能検査をした 20 人（95%）が知的障害と診断され，12 人（63%）は全 IQ 50 以下であった．言語性 IQ と動作性 IQ 間に有意差を認めた（$p<0.0001$）．21 人の K-ABC 施行結果では，継次処理と習得度は同時処理と比較して高値であった．クラスター解析の結果，2 群（A 群 9 例，B 群 12 例）に分けられた．A 群は言語性 IQ，動作性 IQ，全 IQ が B 群より有意に高かった（$p<0.05$）．A 群は継次処理が同時処理と比較して有意に高く，短期聴覚記憶が良好であったが，B 群は継次処理と同時処理の間に有意差はなかった．両クラスター A，B 群間において継次処理のすべての項目と，同時処理の二項目（絵の統合，模様の構成）で有意差を認めた（手の動作，数唱；$p<0.05$，語の配列，絵の統合；$p<0.01$，模様の構成；$p<0.0001$）．一方で「視覚類推」「位置さがし」では両クラスター間で，有意差を認めなかった．

〔結論〕我々の仮説どおり K-ABC で二つのクラスターが同定された．短期的聴覚記憶の良さは WS の全てではなく，一部の WS の特徴といえる．また，空間認知障害が著しいといわれる WS において「同時処理」課題の「位置さがし」課題の低得点は WS に共通して見られ，WS の核となる共通の特徴であることが示唆された一方，「模様の構成」の成績で両群間において有意差を認めたことは，個々の WS の視空間認知機能の障害が様々であることを示唆した．