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Are jigsaw puzzle skills 'spared' in persons with Prader-Willi syndrome?

Elisabeth M. Dykens
University of California, USA

Background: This three-part study examines previous clinical impressions that people with Prader-Willi syndrome have unusual jigsaw puzzle and word search skills. **Results:** Children with Prader-Willi syndrome showed relative strengths on standardized visual-spatial tasks (Object Assembly, Triangles, VMI) in that their scores were significantly higher than age- and IQ-matched peers with mixed mental retardation, but below those of age-matched normal children with average IQs. In striking contrast, children with Prader-Willi syndrome scored on par with normal peers on word searches, and they far *outperformed* them on the jigsaw puzzles, placing more than twice as many pieces as the typically-developing group. Within Prader-Willi syndrome, puzzle proficiency was not predicted by age, IQ, gender, degree of obesity, or obsessive-compulsive symptoms, but by genetic subtypes of this disorder. **Conclusions:** Findings are discussed in relation to splinter skills in autism, and to cases with autism and chromosome 15 anomalies that include the Prader-Willi region. **Keywords:** Behavioral phenotypes, mental retardation, Prader-Willi Syndrome, visuo-spatial functioning.

A flurry of research over the last few years has examined how persons with genetic mental retardation syndromes can inform normal development. Taking advantage of so-called 'experiments of nature' (Hodapp & Burack, 1990), workers have identified how the distinctive abilities and disabilities of people with various disorders – from Williams syndrome to autism – lead to new insights about language, intelligence, and social cognition. Prader-Willi syndrome appears to be another disorder with a jagged profile that may ultimately sharpen theories about cognition, primarily visual-spatial processing.

Prader-Willi syndrome is a genetic disorder associated with developmental disabilities, hyperphagia, and characteristic physical and behavioral features (see Dykens, Hodapp, & Finucane, 2000 for a review). Behaviorally, many persons with Prader-Willi syndrome show persistent food-seeking that, if left unchecked, invariably leads to life-threatening obesity (Dykens, 2000; Holland, Treasure, Coskeran, & Dallow, 1995). Other problems include temper tantrums and obsessive-compulsive features such as skin-picking, hoarding, redoing things, and being overly concerned with symmetry and exactness (Dykens & Kasari, 1997; Dykens, Leckman, & Cassidy, 1996; State, Dykens, Martin, Rosner, & King, 1999).

Relative to maladaptive behavior, however, much less is known about the cognitive features associated with Prader-Willi syndrome (Dykens, 1999). Individuals generally show mild to moderate mental retardation, with relative weaknesses in short-term memory and relative strengths on tasks that assess attention to visual detail, visual-motor coordination, perceptual planning, and spatial organization (Curfs, Weigers, Sommers, Borghgraef, & Fryns, 1991; Dykens, Hodapp, Walsh, & Nash, 1992; Gabel et al., 1986).

Persons with Prader-Willi syndrome are also rumored to have a special talent for assembling jigsaw puzzles. Compared to others with mental retardation, parents report more enjoyment of puzzles in their offspring with Prader-Willi syndrome (Dykens & Rosner, 1999). Based on clinical impressions, Holm and colleagues (1993) included jigsaw puzzle skills as a 'supportive' criterion in the consensus clinical criteria for Prader-Willi syndrome. Puzzle skills may take advantage of the visual-motor strengths shown by many with the syndrome, as well as their obsessive-compulsive tendencies, and needs for order, exactness, and for things to be 'just right' (Dykens et al., 1996; Dykens & Rosner, 1999). Anecdotally, we also observe that many individuals with Prader-Willi syndrome have a fondness for 'word search' puzzles, often carrying word search books with them to school or work.

No studies have yet been published on jigsaw or word search puzzle skills in people with Prader-Willi syndrome, or even in other populations. It thus remains unclear when typically- or atypically-developing children are expected to solve puzzles of varying complexities. Solving a 6-piece puzzle is cast at the 4-year level on the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984), at the 5- to 6-year level on the Battelle Developmental Inventory (Newborg, Stock, Wnek, Guidubaldi, & Svinicki, 1984), and at the 8½-year level on the McCarthy Scales (McCarthy, 1972).

The lack of research on jigsaw puzzles is understandable, as puzzles are not standardized and vary widely in such properties as the number, size, shape, and color of pieces, or the stimuli depicted in

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pictures. In contrast, many standardized tasks are readily available that presumably assess similar visual-motor processes tapped by jigsaw puzzles. Examples include the Object Assembly and Block Design tasks from the Wechsler Intelligence Scale for Children-III (WISC-III; Wechsler, 1991), and the Triangles task from the Kaufman Assessment Battery for Children (K-ABC; Kaufman & Kaufman, 1983). Standardized tasks from IQ tests, however, may somehow differ from the 'unusual skill with jigsaw puzzles' included in the consensus clinical criteria for Prader-Willi syndrome (Holm et al., 1993).

This three-part study assesses how people with Prader-Willi syndrome solve non-standardized jigsaw puzzles and word searches, as well as how they perform on a variety of standardized visual-spatial tasks. As little data exist on puzzle skills in atypical or typical groups, two comparison groups are used. In Study 1, we compare puzzle and visual-spatial skills in children and adolescents with Prader-Willi syndrome to age- and IQ-matched peers with mental retardation of mixed etiologies. In Study 2, we identify if puzzle skills in Prader-Willi syndrome are 'spared', or at chronological age expectations, by comparing children and adolescents with Prader-Willi syndrome to chronological-age matched typically-developing youngsters. In Study 3, we examine possible correlates of puzzle performance in a large cohort of 60 subjects with Prader-Willi syndrome. These include age, gender, IQ, and visual-spatial skills, as well as two variables more specific to Prader-Willi syndrome: obsessive-compulsive tendencies and genetic subtypes of this disorder. Specifically, while a paternally-derived deletion on chromosome 15 (15q11-q13) is found in approximately 70% of cases, about 25% show maternal uniparental disomy (UPD), or when both copies of chromosome 15 are inherited from the mother (see State & Dykens, 2000 for a review). In brief, then, this three-part study examines an area of hypothesized strength, and even possible sparing, in an otherwise cognitively delayed, mentally retarded group.

Study 1

Method

Subjects. This study included 16 people with Prader-Willi syndrome (9 males, 7 females) and 16 age- and IQ-matched people with mental retardation due to heterogeneous etiologies (7 males, 9 females). Subjects in both groups ranged in age from 5 to 25 years; their mean ages are summarized in Table 1. As measured by the Kaufman Brief Intelligence Test (K-BIT; Kaufman & Kaufman, 1990), IQs were similar across the Prader-Willi syndrome and mixed etiology groups (*M*'s = 57.50 and 52.12, respectively), t (30) = 1.02, NS. Among subjects with Prader-Willi syndrome, 12 had genetic testing showing paternal deletions, and 4 had clinical diagnoses (Holm et al., 1993). Among subjects in the mixed group, 3 had Down syndrome, 2 Williams syndrome, 2 autism, 1 cerebral palsy, and 8 unknown reasons for their developmental delay. These types of heterogeneous or mixed groups are widely used in behavioral mental retardation research (Hodapp & Dykens, 1994, 2001).

Procedures

All subjects were individually administered the one-hour test battery by trained research assistants. Mothers completed a demographics questionnaire (child's age, gender, height, weight, diagnostic testing, if appropriate), as well as two other questionnaires described below. The test battery consisted of:

Kaufman Brief Intelligence Test (K-BIT; Kaufman & Kaufman, 1990). The K-BIT assesses cognition in persons aged 4 years through adulthood, and provides standard scores for Vocabulary, Matrices, and a composite IQ. The K-BIT has been successfully used in previous studies of children and adults with mental retardation (e.g., Dykens, Rosner, & Ly, 2001).

Standardized visual-spatial tasks. The Developmental Test of Visual-Motor Integration (VMI; Beery, 1997), a figure copying task, was used to assess written visual-motor functioning. Two tasks, Object Assembly from the WISC-III (Wechsler, 1991), and Triangles from the K-ABC (Kaufman & Kaufman, 1983), were used to assess perceptual organization and visual-motor coordination. In Object Assembly, subjects assemble puzzles depicting four common objects, while in Triangles they assemble yellow and blue triangles to match a stimulus picture. As some subjects' chronological ages exceeded the age range of these tests, raw scores were used in data analyses.

Jigsaw puzzles. The study used two jigsaw puzzles designed specifically for this study; both were made of sturdy cardboard, with 40 pieces per puzzle. Completed puzzles measured 8 × 10 inches, and one puzzle depicted a puppy, the second a slice of pizza. Subjects had no prior experiences with these two puzzles, as the puzzles were novel and created for the study at a local paper goods store.

For each puzzle, subjects were told, 'Now we are going to do some puzzles. I'd like you to try them. Here is what this one looks like'. Subjects were then shown the picture depicted in the puzzle, which was kept in view in front of them. The experimenter then arranged the 40 pieces in random order immediately in front of the subject, such that all pieces were facing upward and with no overlapping pieces. Subjects were told to 'Put the pieces together as fast as you can, and let me know when you are done'. Appropriate encouragement was given to subjects as needed. Raw scores were tabulated by counting the number of pieces that were correctly connected to each other within a three-minute time period. Scores were summed across the two puzzles for a total score that ranged from 0 to 80.

Table 1 Mean ages and IQs, and raw scores (and *t* and *p* values) for visual-spatial and puzzle tasks in the Prader-Willi syndrome versus mixed mental retardation groups

	Prader-Willi		Mixed MR		
	M	*SD*	*M*	*SD*	*t*
Age	14.10	7.93	13.04	5.99	.45
K-BIT IQ	57.50	13.72	52.12	15.97	1.02
Standardized visual-spatial tasks					
Object assembly	14.69	8.08	5.94	5.82	3.51***
Triangles	8.31	5.36	4.77	4.52	2.02*
VMI	13.31	3.16	9.93	4.15	2.59**
Puzzles					
Jigsaw puzzles	30.93	21.35	2.43	2.87	4.95***
Word searches	9.06	6.74	3.44	5.02	2.46**
Parental report					
Puzzle enjoyment	3.81	1.16	2.12	.96	4.47***
Puzzle skill	3.00	1.55	1.07	.27	4.40***
Word search enjoyment	3.06	1.18	1.37	.96	4.99***
Word search skill	1.81	.98	1.18	.40	2.01*

Note. * $p < .05$; ** $p < .01$; *** $p < .001$.

Puzzle strategies. As subjects completed the jigsaw puzzles, the experimenter observed their puzzle-solving strategies on a checklist developed for this study. After informally observing the puzzle-solving strategies of five young adults without mental retardation, we identified six common approaches: does borders first; does center first; looks to picture for guidance; matches colors; tries to force pieces into place; and rotates pieces and attempts to attach at different angles. Prior to data analyses, we reviewed the frequency counts of these strategies across the two puzzles. As there was little variation in strategies across puzzles, analyses used frequency data from the pizza puzzle.

Word searches. Two word searches were designed for this study, and were modeled after approaches used in word search books available in toy and book stores. For each search, 13 words were imbedded at various angles in a 6×6 inch block of letters, with the 13 words listed at the top of the paper. One word search contained common items (e.g., cat, school), while the second included food items (e.g., cookie, peas). For each, subjects were informally asked about their familiarity with word searches, and then told 'Now I want you to look for these words and circle them'. Raw scores were based on the number of words that were circled by subjects within a three-minute time limit. Raw scores were summed into a total word search score that ranged from 0 to 26.

Puzzle and word search enjoyment and skill. Using 1 to 5 scales, parents were asked to answer four questions: 'How much does your child enjoy jigsaw (or word search) puzzles?' (1 = really doesn't like them that much at all to 5 = really loves them); and 'How good is your child at doing jigsaw (or word search) puzzles?' (1 = really fast and accurate to 5 = slower or inaccurate compared to others his/her age).

Yale-Brown Obsessive Compulsive Scale. Parents also completed an informant version of the Y-BOCS (Goodman et al., 1989), which has good reliability and validity (Taylor, 1995). This version contained 30 symptoms rated as being present ever, or in the last week, and data analyses used the sum of lifetime symptoms. Informants also rate the extent to which symptoms are time consuming, distressful, and cause social or adaptive impairment (0 = none to 5 = extreme).

Results

T-tests were used to compare raw scores across groups on Object Assembly, Triangles, and VMI. As shown in Table 1, subjects with Prader-Willi syndrome scored significantly higher than their counterparts with mental retardation on all these tasks.

Table 1 also shows that the group with Prader-Willi syndrome scored significantly higher than the mixed group on both the word searches and jigsaw puzzles; differences were especially striking in the jigsaw puzzles. Indeed, youngsters with Prader-Willi syndrome scored approximately 15 times higher on puzzles than subjects with mixed mental retardation, with 94% of subjects with Prader-Willi syndrome scoring above the highest score in the mixed group. As jigsaw puzzle variances were somewhat large relative to means, we also conducted a rank ordered, non-parametric Mann–Whitney U-test; this also showed a significant group difference in puzzle scores; $z = -2.37$, $p < .01$. Relative to the mixed group, participants with Prader-Willi syndrome had significantly higher parental ratings of jigsaw puzzle and word search enjoyment and skill. As shown in Table 2, Chi-square analyses of the puzzle-solving strategies revealed that relative to the mixed group, those with Prader-Willi syndrome were more apt to

Table 2 Percentage of subjects showing puzzle-solving strategies with the 40-piece pizza puzzle across groups

	Study 1			Study 2		
Strategy	PWS	Mixed	X^2	PWS	Typical	X^2
Borders first	86	10	14.31***	86	50	10.08***
Tries to force	0	40	7.14*	0	29	5.14*
Matches colors	28	0	3.17$^+$	20	14	.20
Rotates pieces	13	11	.06	13	14	.01
Does center first	14	11	.06	06	29	2.68$^+$
Looks to picture	07	11	.09	13	52	5.78**

Note. $^+$ $p < .10$; * $p < .05$; **$p < .01$; *** $p < .001$.

work on the borders first, and less apt to try to force the pieces into place.

Study 2

Method

Subjects. This study included 21 subjects with Prader-Willi syndrome (9 males, 12 females) and 21 typically-developing children and adolescents (8 males, 13 females). Ten of the subjects with Prader-Willi syndrome also participated in Study 1. Seventeen participants had paternal deletions, and 4 had clinical diagnoses (Holm et al., 1993). All subjects ranged in age from 4 to 16 years, and they were individually matched across groups on chronological age. Table 3 summarizes the mean ages and K-BIT IQs from each group; as expected, IQs differed significantly.

Procedures. Test procedures and measures were identical to Study 1.

Results

As shown in Table 3, t-tests revealed that typical children and adolescents scored significantly higher than subjects with Prader-Willi syndrome on Object Assembly, Triangles, and the VMI. However, differences were non-significant on the word searches, with the Prader-Willi group scoring modestly lower than the typical group. In contrast, subjects with Prader-Willi syndrome performed significantly *higher* than the typical group on the jigsaw puzzles (see Table 3). On average, those with Prader-Willi syndrome correctly placed 28.10 pieces, while typically-developing youngsters placed 10.71 pieces. More than twice as many pieces were thus placed by the Prader-Willi syndrome group, and 71% of these subjects earned puzzle scores that were above the typical group's mean score. In contrast, just one subject in the typical group (6%) scored above the Prader-Willi syndrome mean. As with Study 1, puzzle scores were also significant on the non-parametric Mann–Whitney U-test; $z = -2.19$, $p < .05$.

As shown in Table 3, parental reports of skills in jigsaw or word search puzzles were non-significant across groups, though the Prader-Willi group had significantly more enjoyment of puzzles than the typical group. Chi-square analyses of puzzle strategies revealed that relative to typical controls, subjects with Prader-Willi syndrome were more apt to do

Table 3 Mean ages and IQs, and raw scores (and t and p values) for visual-spatial and puzzles tasks in the Prader-Willi syndrome versus typical groups

	Prader-Willi		Typical		
	M	*SD*	*M*	*SD*	*t*
Age	10.60	3.60	9.62	3.27	1.22
K-BIT IQ	63.48	14.75	103.52	10.31	10.20***
Standardized visual-spatial tasks					
Object assembly	14.50	8.98	21.76	12.16	−2.17*
Triangles	7.90	6.16	13.19	4.83	−3.09**
VMI	12.71	4.58	18.47	4.98	−3.90***
Puzzles					
Jigsaw puzzles	28.10	23.65	10.71	9.09	3.14**
Word searches	9.74	7.01	12.43	8.89	−1.05
Parental report					
Puzzle enjoyment	3.90	1.14	2.71	.84	3.85***
Puzzle skill	3.05	1.69	2.57	.81	1.17
Word search enjoyment	2.80	1.70	2.86	1.01	.13
Word search skill	1.90	1.26	2.52	.75	−1.93

Note. * $p < .05$; ** $p < .01$; *** $p < .001$.

the borders first, less apt to look at the picture, and less apt to try to force the pieces into place (see Table 2).

Study 3

Method

Subjects. Sixty (60) individuals with Prader-Willi syndrome participated in the third study, which examined correlates of puzzle performance. Of these, 27 had participated in either Study 1 or Study 2, and 33 had not. Thirty-seven subjects had confirmed paternal deletions, 8 had maternal UPD, and 15 had clinical diagnoses of Prader-Willi syndrome (Holm et al., 1992). Subjects ranged in age from 4 to 38 years, with a mean age of 16.01 years (SD = 8.63). The mean K-BIT IQ of the sample was 64.68 (SD = 14.92). The mean Body Mass Index (BMI; weight in kilos/(height in meters^2) of this group was 27.21 (SD = 9.62), with BMIs that ranged from 14.47 to 55.05. Approximately 65% of the sample were obese, as determined by age-appropriate cut-off points for BMI scores.

Procedures

Test procedures and measures were identical to Study 1 and 2.

Results

We first ensured that the puzzle scores of subjects included in Study 1 and Study 2 were representative of the group of 18, similarly aged persons with Prader-Willi syndrome who had not participated in Study 1 or Study 2. Comparing participants to non-participants of Study 1 or 2, jigsaw puzzle scores were quite consistent; t (58) = .27, NS; M's = 29.52 versus 26.83, respectively. Word search scores, however, were significantly higher in the group that had not previously participated in Study 1 or Study 2; t (58) = −2.42, p < .05; M's = 9.81 versus 15.28, respectively. We may, then, have underestimated word search performance of Prader-Willi subjects in Study 1 and Study 2.

Second, we correlated puzzle performance in the group with Prader-Willi syndrome with age, K-BIT IQ, degree of obesity (BMI), the three standardized visual-spatial tasks, parental reports of puzzle enjoyment and skill, and the Y-BOCS. Table 4 summarizes these correlations. No relationships were found for IQ, Y-BOCS symptoms, or the BMI, nor were there any gender effects. Jigsaw puzzle performance was modestly associated with age, and more strongly associated with the three visual-spatial tasks, and with parental reports of puzzle enjoyment and skill. By way of comparison, similar correlations were also conducted in the typically-developing and mentally retarded control groups. As shown in Table 4, age emerged as a much more robust predictor of puzzle performance in both comparison groups. Relative to the Prader-Willi syndrome group, age correlations were significantly higher in the mixed group, z = 2.43, p = .015, and marginally significantly higher in the typically-developing group, z = 1.87, p = .060.

To further assess relations between visual-spatial tasks and puzzles in participants with Prader-Willi syndrome, two step-wise regression analyses were conducted. Jigsaw puzzle or word search performances were the outcomes, and predictors included age, puzzle enjoyment, K-BIT IQ, Object Assembly, Triangles, and the VMI.

For the jigsaw puzzles in the Prader-Willi syndrome group, parental reports of enjoyment of puzzles emerged as the strongest predictor, accounting for 47% of the variance, F (1, 49) = 38.77, p < .0001. The Object Assembly task captured an additional 7% of the variance, F (2, 48) = 24.94, p < .0001. These two variables thus accounted for 54% of jigsaw puzzle variance. The VMI was the strongest predictor of word searches, accounting for 59% of variance; F (1, 49) = 68.70, p < .001; enjoyment of word searches accounted for an additional 6% of word search variance, F (2, 48) = 42.04, p < .001; while age added 4% of variance, F (3, 47) = 33.12, p < .001. All together, 69% of word search variance was explained by these three variables.

Third, we assessed possible differences in puzzle performance across the two major genetic subtypes of Prader-Willi syndrome. Although our sample of confirmed UPD cases was small, we individually matched the 8 cases with UPD (5 females, 3 males)

Table 4 Correlations between puzzles, subject characteristics and visual-spatial tasks in Prader-Willi syndrome, mentally retarded, and typically-developing groups

	Jigsaw puzzles			Word searches		
	PWS	MR	Typical	PWS	MR	Typical
Age	.33*	.82***	.70***	.52***	.70**	.76***
IQ	−.20	.05	.29	.18	−.16	.09
Triangles	.47***	.48*	.60**	.69***	.52*	.62**
Object assembly	.46***	.72**	.72***	.64***	.50*	.74***
VMI	.48***	.18	.59**	.77***	.44	.65***
Y-BOCS	−.05	.11	.26	−.06	−.23	.27
Enjoyment	.69***	.11	−.03	.68***	.43	.34

Note. * p < .05; ** p < .01; *** p < .001.

on age, gender, and IQ to 8 subjects with paternal deletions. Table 5 summarizes mean ages and IQs for both genetic subtypes. As depicted in Table 5, subjects with UPD scored significantly lower than their counterparts with deletions on the Object Assembly and VMI tasks, and on the jigsaw puzzles. Means on remaining visual-spatial tasks were in the expected direction, but failed to reach significance. Subjects with deletions also had significantly higher parental ratings of puzzle enjoyment and skills.

Discussion

People with Prader-Willi syndrome have relative strengths in visual tasks, but with some highly unusual twists. On one hand, subjects with Prader-Willi syndrome in Study 1 performed significantly better than their age- and IQ-matched peers with mental retardation on the standardized visual-spatial tasks, and on the jigsaw puzzles and word searches. Indeed, children with Prader-Willi syndrome placed 15 times as many puzzle pieces as their mentally retarded counterparts. On the other hand, although Object Assembly, Triangles, and the VMI were relative strengths, they were not spared, falling significantly below the levels achieved by the age-matched group of youngsters without mental retardation in Study 2.

The word search and jigsaw puzzle findings, however, paint a dramatically different picture. On the word searches, children with Prader-Willi syndrome had slightly lower scores than the typical group, suggesting that these skills are at the lower end of the 'normal' range of performance. Yet the jigsaw puzzle findings were the most striking. On these tasks alone, subjects with Prader-Willi syndrome far outperformed the typically-developing group. Thus, despite the lower IQs and mild mental retardation of Prader-Willi syndrome subjects, they successfully placed, on average, more than twice as many jigsaw puzzle pieces as typically-developing youngsters with average IQs.

Such startling findings join only a few other instances of 'sparing' at a group or syndromic level among people with mental retardation. Many persons with Williams syndrome show spared abilities to recognize faces (Wang, Doherty, Rourke, & Bellugi, 1995), as well as the ability to infer complex mental states from eye expressions (Tager-Flusberg, Bishart, & Baron-Cohen, 1998). Expressive language in Williams syndrome was also once thought to be spared (Bellugi, Wang, & Jennigan, 1994), and although such skills are relative strengths compared to overall mental functioning, for the group as a whole they are generally impaired (Mervis, Morris, Bertrand, & Robinson, 1999). While further work is needed, sparing on a group level seems rare.

Why do subjects with Prader-Willi syndrome show high-level performance on the jigsaw puzzles, but only relative strengths on the standardized tasks? Indeed, in regression analyses, Object Assembly explained just 7% of unique jigsaw puzzle variance, which is remarkably low in light of the fact that both tasks are jigsaw puzzles. One could argue that Object Assembly puzzles are more difficult because they do not interlock, are achromatic, and subjects need to figure out what the completed puzzles should be without benefit of a picture or model. But one could also argue that the jigsaw puzzles are harder because they contain many more pieces (40 pieces per puzzle) than the Object Assembly puzzles (M = 6.6 pieces across 5 puzzles). Further, even though a picture was provided for the jigsaw puzzles (and not for Object Assembly), subjects with Prader-Willi syndrome were less apt to refer to the picture than

Table 5 Mean ages, IQs, and raw scores (and t and p values) for visual-spatial and puzzle tasks in 8 subjects with Prader-Willi syndrome due to maternal UPD versus 8 with paternal deletion

	Paternal deletion		Maternal UPD		
	M	*SD*	*M*	*SD*	*t*
Age	14.01	7.55	14.92	9.39	.04
K-BIT IQ	68.43	16.09	64.71	13.85	.06
Standardized visual-spatial tasks					
Object assembly	17.71	8.38	7.83	5.71	−2.47*
Triangles	7.86	7.33	4.33	3.88	−1.55
VMI	14.43	14.50	9.43	5.29	−2.00^{+}
Puzzles					
Jigsaw puzzles	30.43	14.80	4.00	3.85	−4.67***
Word searches	12.43	10.23	7.00	3.94	−1.21
Parental report					
Puzzle enjoyment	4.28	.75	2.00	.89	−4.72***
Puzzle skill	3.50	1.25	1.20	.44	−3.96**
Word search enjoyment	3.16	1.94	2.33	1.96	−.76
Word search skill	2.66	1.86	1.60	.89	−1.17

Note. $^{+}$ $p < .10$; * $p < .05$; ** $p < .01$; *** $p < .001$.

others. Additional studies are needed to identify those properties of puzzles, such as color, or the number and shape of pieces, that facilitate or deter performance.

In a similar vein, the strategies that subjects with Prader-Willi syndrome used to assemble the puzzles need further study. Relative to their counterparts, these individuals were less apt to try to force the pieces into place or use the picture as a reference, and more apt to work on the borders. On first glance, then, it appears that subjects shied away from 'spatial' tactics of rotating pieces and trying to attach them at various angles, and instead may have employed a visual matching approach. Such preliminary observations warrant further study, as they touch on the two primary types of visual information processing. Specifically, the visual memory system appears to retain information about static visual features or patterns (and may be associated with the ventral region of the posterior cortex), while the spatial memory system retains dynamic information about movement and movement sequences (and may be associated with the superior parietal lobe) (Logie & Pearson, 1997; Postle & D'Esposito, 1999; Postle, Zarahan, & D'Esposito, 2000). It is not clear which of these visual systems are primarily enlisted by persons with Prader-Willi syndrome as they solve jigsaw puzzles, and how their approach might compare to other advanced-level jigsaw puzzlers without this disorder.

Genetic explanations for the advanced-level puzzle performance in persons with Prader-Willi syndrome remain also elusive. While a number of imprinted and nonimprinted genes have now been identified in the Prader-Willi/Angelman syndrome critical region on chromosome 15, it is unclear how they relate to the Prader-Willi behavioral phenotype (Cassidy, Dykens, & Williams, 2000). Although the genetic mechanisms are as yet unknown, Study 3 provides a first look at other possible correlates of puzzle skills in a large cohort of 60 individuals. Jigsaw puzzle performance was not significantly related to IQ, nor to the obsessive-compulsive features that characterize Prader-Willi syndrome. Although some individuals with Prader-Willi syndrome might develop an obsessive-like interest in puzzles, their obsessions and compulsions in general do not seem predictive of jigsaw puzzle or word search proficiency.

Although age was modestly correlated with puzzle performance, it did not emerge as a significant predictor of jigsaw puzzle variance for participants with Prader-Willi syndrome. Age, however, was strongly correlated with puzzle performance in both the typically-developing group and among participants with mixed mental retardation. Theoretically, advancing age should bring about increased exposure to puzzles, and more opportunities to practice with them. Yet even relatively young children with Prader-Willi syndrome in this study may have been 'practicing' quite a bit, thereby lowering this correlation in this group. Indeed, our top three puzzle 'stars' (completing 62, 66, and 69 pieces) were all young, and their ages of 9, 13, and 15 years fell below the mean age of the large Prader-Willi syndrome sample (16.04 years).

Although puzzle proficiency does not seem closely tied to age in Prader-Willi syndrome, a limitation of this study is that we did not directly assess exposure to puzzles or practice with them. It is thus possible that children with Prader-Willi syndrome have simply had more practice with puzzles, and are thus more proficient than their typically-developing or mentally retarded peers. Furthermore, individual differences in practice or exposure may help explain within-syndrome variability, and why not all children with Prader-Willi syndrome are proficient with puzzles. Despite possible differences in practice, however, parents of Prader-Willi syndrome and typically-developing children reported equal levels of puzzle skills in their offspring, but more enjoyment of puzzles in the Prader-Willi syndrome group. Indeed, enjoyment of puzzles emerged as the strongest predictor of puzzle performance in the Prader-Willi syndrome group, and in regression analyses, accounted for 47% of jigsaw puzzle variance.

But even if pleasure or practice with puzzles contributes to puzzle proficiency, these explanations beg the question of why children with Prader-Willi syndrome are attracted to jigsaw puzzles to begin with (enough so for puzzles to be included in the consensus clinical criteria for this disorder). Although the reasons are presently unknown, certain aspects of puzzle skills in Prader-Willi syndrome have a savant-like quality. Hill (1978) notes that savant syndrome applies to a 'mentally retarded person demonstrating one or more skills *above* the level expected of non-retarded individuals' (p. 281). Technically, the superior performance of the Prader-Willi group meets this definition. Yet savant syndrome is quite rare; as many as 50% of those with savant syndrome have autism, and just five or so types of savant syndrome have been observed over the years (i.e., calendar calculating, music, rote memory, drawing, mechanical). Finally, savant skills are unattainable by most people in the general population; certainly being good at puzzles is within reach of many people.

Instead of savant syndrome, the advanced puzzle skills of persons with Prader-Willi syndrome more closely resemble 'splinter skills', especially as they are seen in persons with autism (Prior, 1979). These circumscribed skills are above the individual's general intellectual level, but would not be considered remarkable or extraordinary in the absence of mental retardation (Nettelbeck, 1999). Common splinter skills in persons with autism are elevated performances on visual-spatial and pattern recognition tasks, including on Block Design, Object Assembly, and the Imbedded Figures Test (Shah & Frith, 1983, 1993; Jolliffe & Baron-Cohen, 1997).

Although formal studies are lacking, many persons with autism also excel at jigsaw puzzles (Prior, 1979).

Such observations are particularly intriguing in light of recent connections between autism and chromosome 15 anomalies, especially in the region of 15q11, which encompasses the Prader-Willi/Angelman syndrome region. In particular, children with isodicentric chromosome 15, previously known as inverted duplication, often show autistic symptoms or full-blown autistic disorder (Martin et al., 2000; Rineer, Finucane, & Simon, 1998; Wolpert et al., 2000). All cases of isodicentric chromosome 15 reported by Schroer et al. (1998) and Wolpert et al. (2000) were of maternal origin. These persons have the characteristic symptoms of autism, but their neuropsychological profiles have not been reported, including whether they show visual-spatial strengths or weaknesses.

It is also unknown how the lack of paternally-derived imprinted information to the Prader-Willi region on chromosome 15 is associated with puzzle skills, or why they vary across genetic subtypes of Prader-Willi syndrome. Although the sample size was small, subjects with maternal UPD performed much worse than their matched counterparts with paternal deletions on the visual-spatial tasks and jigsaw puzzles. Dykens, Cassidy, and King (1999) found that persons with UPD had higher IQs (primarily Verbal IQs, see Roof et al., 2000), as well as less severe or frequent problems such as skin-picking, hoarding, and overeating. Yet those with UPD may also have a reduced capacity to discriminate forms that require the use of stereoscopic vision (Roof et al., 1999), which is likely associated with the poor puzzle performance of the maternal UPD cases in the present study. Thus, compared to those with paternal deletions, those with maternal UPD may have poorer visual and/or visual-spatial processing abilities, but slightly spared verbal skills and less behavioral dysfunction. The genetic mechanisms for these phenotypic differences across genetic subtypes remain unknown. Possible explanations include: incomplete or leaky imprinting (leading to a partial or low level of expression of genes in two doses in UPD cases but only one dose in deletion cases); haploinsufficiency of nonimprinted genes in cases with paternal deletions, or an overexpression of some gene(s) in persons with maternal UPD (Cassidy et al., 2000).

As the first study to identify a sparing in jigsaw puzzles (and possibly word searches) in paternally deleted cases of Prader-Willi syndrome, this study raises more questions than it answers. Future studies are needed on possible genetic, neurological, and environmental reasons for such advanced skills in people with an otherwise profoundly debilitating disorder. Ultimately, this work may shed new light on visual versus spatial information processing among persons in general, or the development of visual or spatial skills in typically-developing children without Prader-Willi syndrome. In these ways, these unusual findings from Prader-Willi syndrome, as with those from Williams syndrome, autism, and other disorders, underscore the potential of using abnormal groups to inform normal development.

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Correspondence to

Elisabeth M. Dykens, University of California, Neuropsychiatric Institute, 760 Westwood Plaza, Los Angeles, CA 90024, USA; Email: edykens@mednet.ucla.edu

References

Beery, K.E. (1997). *Developmental test of visual-motor integration* (3rd revision). Cleveland, OH: Modern Curriculum Press.

Bellugi, U., Wang, P., & Jernigan, T.L. (1994). Williams syndrome: An unusual neuropsychological profile. In S.H. Browman & J. Grafram (Eds.), *Atypical cognitive deficits in developmental disorders* (pp. 23–56). Hillsdale, NJ: Erlbaum.

Cassidy, S.B., Dykens, E.M., & Williams, C.A. (2000). Prader-Willi and Angelman syndromes: Sister imprinted disorders. *American Journal of Medical Genetics*, *97*, 136–146.

Curfs, L.G., Wiegers, A.M., Sommers, J.R., Borghgraef, M., & Fryns, J.P. (1991). Strengths and weaknesses in the cognitive profile of youngsters with Prader-Willi syndrome. *Clinical Genetics*, *40*, 430–434.

Dykens, E.M. (1999). Prader-Willi syndrome: Toward a behavioral phenotype. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders* (pp. 137–154). Cambridge, MA: MIT Press.

Dykens, E.M. (2000). Contaminated and unusual food combinations: What do people with Prader-Willi syndrome choose? *Mental Retardation*, *38*, 163–171.

Dykens, E.M., Cassidy, S.B., & King, B.H. (1999). Maladaptive behavior differences in Prader-Willi syndrome due to paternal deletion versus maternal uniparental disomy. *American Journal on Mental Retardation*, *104*, 67–77.

Dykens, E.M., Hodapp, R.M., & Finucane, B.M. (2000). *Genetics and mental retardation syndromes: A new look at behavior and interventions*. Baltimore, MD: Brookes.

Dykens, E.M., Hodapp, R.M., Walsh, K.K., & Nash, L. (1992). Profiles, correlates, and trajectories of intelligence in Prader-Willi syndrome. *Journal of the Academy of Child and Adolescent Psychiatry*, *31*, 1125–1130.

Dykens, E.M., & Kasari, C. (1997). Maladaptive behavior in children with Prader-Willi syndrome, Down syndrome, and non-specific mental retardation. *American Journal on Mental Retardation*, *102*, 228–237.

Dykens, E.M., Leckman, J.F., & Cassidy, S.B. (1996). Obsessions and compulsions in Prader-Willi syndrome. *Journal of Child Psychology and Psychiatry*, *37*, 995–1002.

Dykens, E.M., & Rosner, B.A. (1999). Refining behavioral phenotypes: Personality-motivation in Williams and Prader-Willi syndromes. *American Journal on Mental Retardation*, *104*, 158–169.

Dykens, E.M., Rosner, B.A., & Ly, T.M. (2001). Drawings by individuals with Williams syndrome: Are people different from shapes? *American Journal on Mental Retardation*, *106*, 94–107.

Gabel, S., Tarter, R.E., Gavaler, J., Golden, W., Hegedus, A.M., & Mair, B. (1986). Neuropsychological capacity of Prader-Willi children: General and specific aspects of impairment. *Applied Research in Mental Retardation*, *7*, 459–466.

Goodman, W.K., Price, L.H., Rasmussen, S.A., Mazure, C., Fleischmann, R.L., Hill, C.L., Heninger, G.R., & Charney, D.S. (1989). The Yale-Brown Obsessive-Compulsive Scale: Development, use, and reliability, *Archives of General Psychiatry*, *46*, 1006–1011.

Hill, A. (1978). Savants: Mentally retarded individuals with special skills. In N. Ellis (Ed.), *International review of research in mental retardation* (pp. 277–297). New York: Academic Press.

Hodapp, R.M., & Burack, J.A. (1990). What mental retardation teaches us about typical development: The examples of sequences, rates, and cross-domain relationships. *Developmental Psychopathology*, *2*, 213–225.

Hodapp, R.M., & Dykens, E.M. (1994). Mental retardation's two cultures of behavioral research. *American Journal on Mental Retardation*, *98*, 675–687.

Hodapp, R.M., & Dykens, E.M. (2001). Strengthening behavioral research on mental retardation syndromes. *American Journal on Mental Retardation*, *106*, 4–15.

Holland, A.J., Treasure, J., Coskeran, P., & Dallow, J. (1995). Characteristics of the eating disorder in Prader-Willi syndrome: Implications for treatment. *Journal of Intellectual Disability Research*, *39*, 373–381.

Holm, V.A., Cassidy, S.B., Butler, M.G., Hanchett, J.M., Greenswag, L.R., Whitman, B.Y., & Greenberg, F. (1993). Prader-Willi syndrome: Consensus diagnostic criteria. *Pediatrics*, *91*, 398–402.

Jolliffe, T., & Baron-Cohen, S. (1997). Are people with Asperger syndrome faster than normal on the Imbedded Figures Test? *Journal of Child Psychology and Psychiatry*, *38*, 527–534.

Kaufman, A.S., & Kaufman, N.L. (1983). *Kaufman Assessment Battery for Children*. Circle Pines, MN: American Guidance Service.

Kaufman, A.S., & Kaufman, N.L. (1990). *Kaufman Brief Intelligence Test*. Circle Pines, MN: American Guidance Service.

Logie, R.H., & Pearson, D.G. (1997). The inner eye and the inner scribe of visuo-spatial working memory: Evidence from developmental fractionation. *European Journal of Cognitive Psychology*, *9*, 241–257.

Martin, E.R., Menold, M.M., Wolpert, C.M., Bass, M.P., Donnelly, S.L., Ravan, S.A., Zimmerman, A., Gilbert, J.R., Vance, J.M., Maddox, L.O., Wright, H.H., Abramson, R.K., DeLong, G.R., Cuccaro, M.L., & Perick-Vance, M.A. (2000). Analysis of linkage disequilibrium in gamma-amniobutyric acid receptor subunit genes in autistic disorder. *American Journal of Medical Genetics*, *96*, 43–48.

McCarthy, D.A. (1972). *Manual for the McCarthy Scales of Children's Abilities*. San Antonio: The Psychological Corporation.

Mervis, C.B., Morris, C.A., Bertrand, J.M, & Robinson, B.F. (1999). Williams syndrome: Findings from an integrated program of research. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders* (pp. 65–110). Cambridge, MA: MIT Press.

Nettelbeck, T. (1999). Savant syndrome-rhyme without reason. In M. Anderson (Ed.), *The development of intelligence* (pp. 247–274). East Sussex: Psychology Press.

Newborg. J., Stock, J.R., Wnek, L., Guidubaldi, J., & Svinicki, J. (1984). *The Battelle developmental inventory*. Allen, TX: DLM/Teaching Resources.

Postle, B.R., & D'Esposito, M. (1999). 'What—then—where' in visual working memory: An event-related fMRI study. *Journal of Cognitive Neuroscience*, *11*, 585–597.

Postle, B.R., Zarahn, E., & D'Esposito, M. (2000). Using even-related fMRI to assess delay-period activity during performance of spatial and nonspatial working memory tasks. *Brain Research Protocols*, *5*, 57–66.

Prior, M. (1979). Cognitive abilities and disabilities in autism: A review. *Journal of Abnormal Child Psychology*, *2*, 357–380.

Rineer, S., Finucane, B.M., & Simon, E.W. (1998). Autistic symptoms among children and young adults with isodicentric chromosome 15. *American Journal of Medical Genetics, 81*, 428–433.

Roof, E., Fox, R., Feurer, I.D., Stone, W., MacLean, W., Butler, M.G., & Thompson, T. (1999, July). *Visual perception in Prader-Willi syndrome*. Paper presented to the 14th Annual Prader-Willi Syndrome Conference, San Diego, CA.

Roof, E., Stone, W., MacLean, W., Feurer, I.D., Thompson, T., & Butler, M.G. (2000). Intellectual characteristics of Prader-Willi syndrome: Comparison of genetic subtypes. *Journal of Intellectual Disability Research, 44*, 25–30.

Schroer, R.J., Phelan, M.C., Michaelis, R.C., Crawford, E.C., Skinner, S.A., Cuccaro, M., Simeensen, R.J., Bishop, J., Skinner, C., Fender, D., & Stevenson, R.E. (1998). Autism and maternally derived aberrations of chromosome 15. *American Journal of Medical Genetics*, *76*, 327–336.

Shah, A., & Frith, U. (1983). An islet of ability in autistic children: A research note. *Journal of Child Psychology and Psychiatry*, *24*, 613–620.

Shah, A., & Frith, U. (1993). Why do autistic individuals show superior performance on the Block Design task? *Journal of Child Psychology and Psychiatry*, *34*, 1351–1364.

Sparrow, S.S., Balla, D., & Cicchetti, D.V. (1984). *Vineland Adaptive Behavior Scales*. Circle Pines, MN: American Guidance Service.

State, M.W., & Dykens, E.M. (2000). Prader-Willi syndrome: Genes, brain, and behavior. *Journal of the American Academy of Child and Adolescent Psychiatry*, *39*, 797–800.

State, M.W., Dykens, E.M., Rosner, B., Martin, A., & King, B.H. (1999). Obsessive-compulsive symptoms in Prader-Willi and 'Prader-Willi-like' patients. *Journal of the American Academy of Child and Adolescent Psychiatry*, *38*, 329–334.

Tager-Flusberg, H., Boshat, J., & Baron-Cohen, S. (1998). Reading the windows to the soul: Evidence of domain-specific sparing in Williams syndrome. *Journal of Cognitive Neuroscience*, *10*, 631–639.

Taylor, S. (1995). Assessment of obsessions and compulsions: reliability, validity, and sensitivity to treatment effects. *Clinical Psychology Review, 15*, 261–296.

Wang, P.P, Doherty, S., Rourke, S.B., & Bellugi, U. (1995). Unique profile of visuo-perceptual skills in a genetic syndrome. *Brain and Cognition*, *29*, 54–65.

Wechsler, D. (1991). *Manual for the Wechsler Intelligence Scale for Children – Third Edition*. San Antonio, TX: The Psychological Corporation.

Wolpert, C., Menold, M.M., Bass, M.P., Qusiyeh, M.B., Donnelly, S.L., Ravan, S.A., Vance, J.M., Gilbert, J.R., Abramson, R.K., Wright, H.H., Cuccaro, M.L., & Pericak-Vance, M.A. (2000). Three probands with autistic disorder and isodicentric chromosome 15. *American Journal of Medical Genetics*, *96*, 365–372.

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