

Reading the Windows to the Soul: Evidence of Domain-Specific Sparing in Williams Syndrome

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Abstract

■ This study tested the hypothesis that Williams syndrome, a rare genetic neurodevelopmental disorder with an unusual cognitive phenotype, involves spared abilities in the domain of understanding other minds. A group of retarded adults with Williams syndrome was compared to an age-, IQ-, and language-matched group of adults with Prader-Willi syndrome, another genetic disorder without the cognitive characteristics of Williams syndrome, and a group of age-matched normal adults, on a task that taps mentalizing ability. The task involved selecting the correct labels to match photographs of complex mental state expressions in the eye region of the face. The adults with

Williams syndrome performed significantly better than the adults with Prader-Willi on this task, and about half the group performed in the same range as the normal adults. These findings are consistent with anecdotal evidence about Williams syndrome and provide evidence that mentalizing is a distinct cognitive domain. This spared cognitive capacity may be linked to the relative sparing of limbic-cerebellar neural substrate in Williams syndrome, which is also connected to cortico-frontal regions that are known to be involved in understanding complex mental states. ■

INTRODUCTION

Individuals with Williams syndrome have a unique cognitive-behavioral phenotype that includes relative sparing in the domains of language and face processing in the presence of marked deficits in visual-spatial cognition and mild to moderate levels of mental retardation (Bellugi, Marks, Bihrlé, & Sabo, 1988; Bellugi, Bihrlé, Neville, Jernigan, & Doherty, 1992; Mervis & Bertrand, 1997). Furthermore, they are socially motivated, with a strong interest in other people, coupled with a warm and friendly personal style (Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987). Indeed, infants and children with Williams syndrome are unusually attentive to faces (Mervis & Bertrand, 1997) and have been described as very empathic toward other people (Gosch & Pankau, 1994). This profile has led some researchers to propose that Williams syndrome may be characterized by relative sparing in understanding other minds (Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995; Tager-Flusberg & Sullivan, 1996). If Williams syndrome does involve relative sparing in this domain of understanding minds, or *mentalizing*, this neurodevelopmental disorder would stand in striking contrast to autism, another neurodevelopmental disorder that has been clearly char-

acterized as involving specific impairment in this cognitive domain (Baron-Cohen, Leslie, & Frith, 1985; Baron-Cohen, Tager-Flusberg, & Cohen, 1993). This kind of dissociation would provide strong evidence that the capacity for mentalizing represents a distinct cognitive domain with an associated neural substrate that may be uniquely affected by a neurodevelopmental disorder of genetic origin, relatively independent of general cognitive or language capacities.

Williams syndrome is a rare genetic disorder that is caused by a submicroscopic hemizygous contiguous gene deletion on chromosome 7 in the q11.32 region. Thus far two of the genes that are involved in Williams syndrome have been identified: the elastin gene, which is responsible for the heart and other connective or soft tissue disorders that are typical in this syndrome (Ewart et al., 1993; Morris et al., 1994), and LIM-kinase 1, which is contiguous with the elastin gene and accounts for the visuospatial constructive deficit that characterizes the most striking feature of the cognitive impairment in Williams syndrome (Frangiskakis et al., 1996). At this point, the genetic source of the unusual social behavior associated with the Williams syndrome phenotype has not been identified, although it is clear that additional genes are involved.

At the neurobiological level, individuals with Williams syndrome have significantly smaller overall brain volumes compared to normal, with significant reductions in total cerebral gray matter (Jernigan & Bellugi, 1990, 1994; Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993). However, there are no significant differences between Williams syndrome and normal controls in neocerebellar volume and limbic structures, including the amygdala hippocampus, parahippocampal gyrus, and uncus (Bellugi, Wang, & Jernigan, 1994; Galaburda, Wang, Bellugi, & Rossen, 1994). Compared to Down syndrome subjects, the anterior cortical regions are significantly larger in volume. These anatomical regions that show specific sparing in Williams syndrome, particularly the neocerebellum and the limbic system, have been implicated as sites of neuropathology in autism in a range of studies (Bauman & Kemper, 1994; Courchesne et al., 1994; Minshew, Sweeney, & Furman, 1995).

Studies of nonhuman primates and adults with brain damage have suggested that the processing of social information, particularly faces and affective expressions, can be linked to the limbic system, especially the amygdala and associated medial temporal cortical structures (Baron-Cohen & Ring, 1994; Brothers & Ring, 1992; Brothers, Ring, & Kling, 1990; Eslinger & Damasio, 1985; Perrett et al., 1990), and to frontal regions that are directly connected to the neocerebellum-limbic complex (Baron-Cohen, 1995; Price, Russchen, & Amaral, 1987). Taken together, the neurobiological evidence suggests that these regions may be specifically implicated in the relatively spared social cognitive abilities in Williams syndrome.

There is, however, little beyond anecdotal evidence for spared mentalizing abilities in Williams syndrome. Only one study has systematically investigated theory of mind performance in Williams syndrome (Karmiloff-Smith et al., 1995), using task paradigms that have been employed in developmental psychological research. This study provided evidence that the majority of individuals with Williams syndrome do pass first-order theory of mind tasks, such as false belief, and that some even pass second-order tasks (such as attributing second-order beliefs and interpreting nonliteral language). However, because of methodological limitations in this study, it does not provide clear-cut evidence for sparing in this population in this domain. First, the subjects with Williams syndrome ranged in age between 9 and 23, well beyond the age at which normal children pass the first- and second-order theory of mind tasks that were used. Second, an appropriate control group was not included; Karmiloff-Smith et al. (1995) only compared the performance of their Williams syndrome subjects to data from autistic subjects, who are known to be specifically impaired on theory of mind tasks. It is important to include some kind of appropriate comparison group in studies of this sort because most individuals with Williams syndrome are mentally retarded. Third, most of the tasks used by

Karmiloff-Smith and her colleagues were language-based, requiring the subjects to follow detailed narratives and to answer grammatically complex questions. Perhaps the relatively good performance of the Williams syndrome subjects had more to do with their relatively spared language rather than spared theory of mind. Thus, it is not clear from this study whether children with Williams syndrome perform significantly better than age-, language-, and IQ-matched children, who may very well pass these same tasks. The inclusion of an appropriate and well-matched comparison group would therefore be needed to provide the critical test for spared mentalizing ability in this population.

In this study, we attempt to address some of these methodological concerns, while providing a direct test of the hypothesis that individuals with Williams syndrome have a relatively spared mentalizing ability. The task we employed was developed by Baron-Cohen and his colleagues for use with adults (Baron-Cohen, Jolliffe, Mortimore, & Robertson, 1997; see also Baron-Cohen, 1995) and is referred to as the "Eyes task." It directly measures a subject's ability to interpret the expression of a range of mental states in the eye region of the face, extending an earlier idea of Numenmaa (1964) that there might be a "language of the face" for expressing mental states. Baron-Cohen, Jolliffe, et al. (1997) argue that this task should be considered a test of "mindreading," or mentalizing ability, because the subject has to choose the appropriate mental state label to match the expression in the eyes. Furthermore, the task has no executive function components and therefore represents a fairly pure measure of mentalizing ability. The task does, however, test the subjects' ability to attribute a full range of mental states, not just basic emotions. One advantage of using this task for our study is that it does not involve tracking complex narratives, as do all other more advanced-level theory of mind tasks (e.g., Happé, 1994). Because it has been developed for use with adults, it is age appropriate for the subjects in our study. Furthermore, the task is a more sensitive measure for differences between individuals and groups because it is not limited to a pass/fail scoring system and is less likely to produce ceiling effects. Not surprisingly, high-functioning adults with autism perform poorly on this task even though they pass control tasks requiring them to recognize basic emotions and identify gender differences (Baron-Cohen, Jolliffe, et al., 1997; Baron-Cohen, Wheelwright, & Jolliffe, 1997). These findings are consistent with what is known about the specific mentalizing deficits in autism. Baron-Cohen and his colleagues have also used this task to reveal small but significant differences between parents and siblings of people with autism or Asperger syndrome in comparison to individuals who do not have close family members with either of these disorders and in comparison to family members of children with Tourette syndrome (Baron-Cohen & Hammer, 1997).

In the present study, we compare the performance of

adults with Williams syndrome on the Eyes task with adults with Prader-Willi syndrome, another well-defined genetic disorder caused by a loss of paternal genetic information on chromosome 15, in the q11-q13 region (Butler, 1990, 1994; Ledbetter et al., 1980). The loss may be the result of paternal deletion, uniparental maternal disomy, or an imprinting mutation (Nicholls, Knoll, Butler, Karam, & Lalonde, 1989). Although people with Prader-Willi syndrome have a similar IQ distribution to individuals with Williams syndrome, their cognitive profile is more balanced (Dykens, in press; Thompson, Butler, MacLean, Joseph, & Delaney, in press). Thus Prader-Willi syndrome does not appear to involve a consistent uneven cognitive phenotype, and language ability tends to be similar to overall cognitive level. Furthermore, the social-affective behavior of children and adults with Prader-Willi tends to be similar to other retarded individuals at the same cognitive level, in contrast to either Williams syndrome or autism. Thus Prader-Willi syndrome provides an excellent comparison group for our Williams syndrome subjects because they can be easily matched to them on age, IQ, and language ability. In this way we could test whether adults with Williams syndrome are significantly better than matched retarded subjects in the domain of mentalizing, which would suggest relative sparing of function. We also included a group of normal age-matched adults in order to test the more stringent hypothesis that Williams syndrome may involve *absolute* sparing of mindreading ability, despite their mild to moderate levels of mental retardation.

RESULTS

Table 1 shows the mean scores and standard deviations on the Eyes task for each group. In contrast to findings reported by Baron-Cohen, Jolliffe, et al. (1997), an initial analysis of variance (ANOVA) revealed no sex differences in the number of correct responses to the Eyes task ($F(1, 49) = 0.12, ns$). This variable, therefore, was omitted from any further analysis. Perhaps because our sample was relatively small, we lacked the power to detect the sex differences found by Baron-Cohen, Jolliffe, et al. (1997). A one-way ANOVA revealed significant group effects ($F(2, 48) = 12.00, p < 0.0001$). A post hoc t test indicated that the normal group performed significantly better than the Prader-Willi syndrome group (critical difference = 2.61,

$p < 0.01$) and the Williams syndrome group (critical difference = 2.16, $p < 0.05$) and that the Williams syndrome group performed significantly better than the Prader-Willi syndrome group (critical difference = 2.16, $p < 0.05$).

It is important to notice that although the Williams syndrome group scored significantly better on the Eyes task than the Prader-Willi syndrome group, they did not perform at quite the same levels as the normal group. Although the two clinical groups were matched on receptive vocabulary and IQ, the means for Williams syndrome adults on these standardized measures were slightly higher than the means for the Prader-Willi syndrome subjects. Because of the small sample size we could not use these measures as covariates in our analyses, so we therefore chose to follow up with nonparametric analyses of the data from the two clinical groups to confirm that the Williams syndrome subjects were significantly better on the task.

Using binomial theorem, scores of 17 or more out of 25 were significantly above chance (assuming chance on each trial at 0.5) and considered a *passing score*. All scores lower than 17 were considered to be at chance. Looking just at the means for the groups, we see that both the normal and Williams syndrome groups can be viewed as passing this task, whereas the Prader-Willi group as a whole did not perform above chance. At an individual subject level, among the normal adults 23 of 25 passed the task, 8 of 13 Williams syndrome adults passed, and only 3 of 13 Prader-Willi syndrome adults passed. The difference in the number of passers between the Williams syndrome and Prader-Willi syndrome groups was significant ($s(\chi^2 = 3.94, p < 0.05)$). Table 2 shows the mean IQ and Peabody Picture Vocabulary Test-Revised (PPVT-R) mental age for the passers and those who were performing at chance in each group. Within each group there were no significant IQ or PPVT mental age differences on the Eyes task between the passers and those who did not score significantly above chance.

If we omit the two normal adults who performed at chance, the range of scores for this group was between 18 and 23. Six of 13 adults with Williams syndrome scored within this range compared to only 2 of 13 adults with Prader-Willi syndrome.

To examine the possibility that vocabulary difficulties led to reduced performance in the clinical groups on

Table 1. Performance on the Eyes Task

	Group		
	Normal	Williams Syndrome	Prader-Willi Syndrome
Mean score (out of 25)	19.6	17.3	14.8
Standard deviation	2.2	3.8	3.1

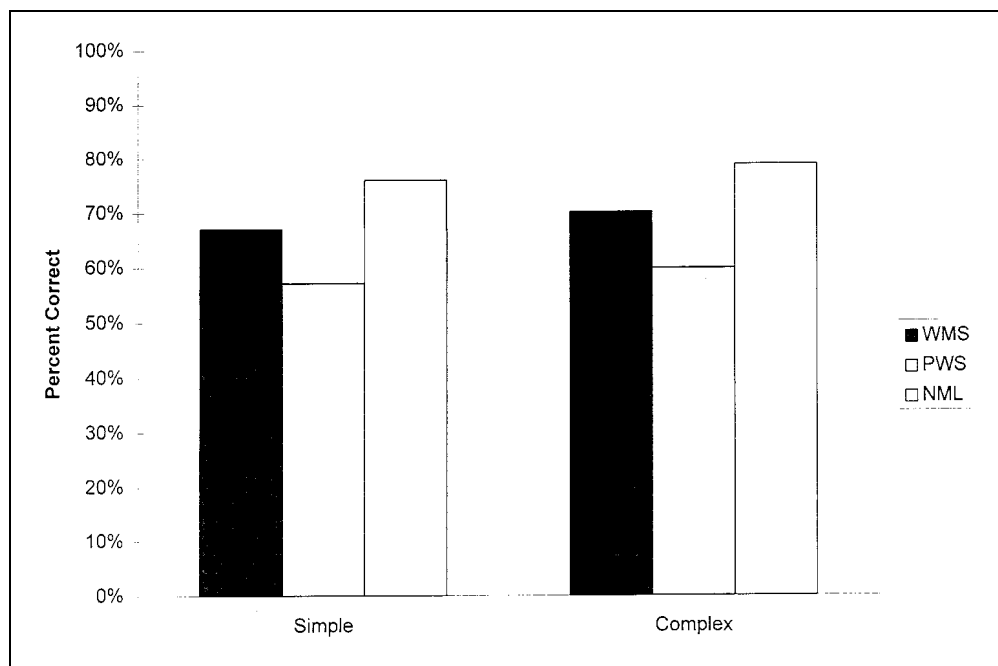
Table 2. Characteristics of Subjects Who Pass or Perform at Chance on the Eyes Task

	Group			
	Williams Syndrome		Prader-Willi Syndrome	
	Pass	Chance	Pass	Chance
N	8	5	3	10
IQ composite score	63.8 (9.5)	61.2 (11.1)	59.0 (4.0)	56.0 (11.1)
PPVT-R mental age	13;8 (8;4)	11;5 (2;2)	12;4 (3;4)	8;7 (1;10)

this task, we examined how the three groups responded on individual items. If vocabulary was a factor, certain items should be difficult across groups, especially for the clinical groups, and those items should tend to involve more difficult vocabulary. Percentages of subjects in each group responding incorrectly were computed for each item. Correlations were then computed to see if there were consistent patterns among the three groups. The response patterns for the Williams syndrome and Prader-Willi syndrome groups were significantly correlated ($r(25) = 0.50, p < 0.01$). Interestingly, although the response patterns for the Prader-Willi syndrome and normal groups were not all similar ($r(25) = 0.12, ns$), the correlation between the Williams syndrome and normal groups did almost reach significance ($r(25) = 0.35, p < 0.09$). Thus, the adults with Williams syndrome performed in quite similar ways to both the normal adults and the adults with Prader-Willi syndrome. Examination of the individual items showed that the items that the Prader-Willi syndrome and Williams syndrome found

more difficult (i.e., made more errors on) were not necessarily those with more difficult vocabulary terms associated with them. For example, on the item “Sad thought/Happy thought,” no normal adults answered incorrectly, whereas 23% of Williams syndrome adults and 62% of Prader-Willi syndrome adults answered incorrectly. In contrast, on the item “Sympathetic/Unsympathetic,” 60% of normal adults answered incorrectly, 30% of the Williams syndrome adults answered incorrectly, and only 8% of the Prader-Willi syndrome adults answered incorrectly.

It was possible that for some items, subjects could choose the correct item based just on the direction of the gaze of the eyes (e.g., “Noticing you/Ignoring you”). To rule out this possibility, we divided the items into 7 “simple” items, or those which could be answered based on eye gaze alone (Items 2, 6, 15, 18, 19, 20, 21; see “Appendix”), and 18 “complex” items, which could not. As can be seen in Figure 1, subjects performed similarly on the simple and complex items.

Figure 1. Percentage correct, by group and complexity.

DISCUSSION

The main finding from this study is that adults with Williams syndrome perform significantly better than an age-, IQ-, and language-matched group of retarded individuals in their performance on a task that directly taps the ability to attribute mental states to other people. At the same time, because the Williams syndrome group as a whole did not perform on this task as well as age-matched normally intelligent adults, we cannot conclude that this ability is spared in an absolute sense in this population, although almost half the Williams syndrome subjects did perform in the same range as our normal controls, and their response profile was similar. Given the lower IQ scores of our Williams syndrome sample, it is perhaps not surprising that some subjects did not perform very well; presumably attentional and memory factors in this group would limit their ability on any structured task.

The relative sparing of this kind of mentalizing capacity in Williams syndrome cannot simply be attributed to IQ or language ability alone because the two clinical groups were matched on these variables. Furthermore, we did not find item effects that would have indicated that some vocabulary terms were more difficult than others, and there were no language or IQ differences between those subjects who passed the task and those who performed at chance. This suggests that the ability to read mental state information from eyes is a cognitive domain that is somewhat independent of general cognitive abilities and language in these populations.

The findings from this study need to be replicated using additional measures of mentalizing ability. Although the Eyes task has proven successful in studies in identifying subtle deficits among individuals with autism and Asperger syndrome and their family members (Baron-Cohen & Hammer, 1997; Baron-Cohen, Jolliffe, et al., 1997), there are some limitations to the task. One concern is that the target items have not been counter-balanced with the foil items; for each pair of eyes only one target term is ever the correct choice. Furthermore, there are slightly more positively valenced target terms (e.g., *concerned*) than negatively valenced terms (e.g., *unconcerned*). Although there may have been some subtle bias in the task that favored the Williams syndrome group, we should note that the adults with Williams syndrome did not perform differently across these two groups of target terms.

The findings from this study suggest that in Williams syndrome there is selective sparing of the cognitive capacity referred to here as mentalizing ability. Clearly adults with Williams syndrome are quite good at reading both simple and more complex mental state information from the eye region of the face. This fits with reports of their sensitivity to the feelings of other people and with their general interest in people. It is not clear, however, that this kind of ability reflects more broadly defined

theory of mind capacities. Based on this study we do not know whether Williams syndrome involves relative sparing in understanding the mind as a representational system because studies using tasks that tap this more specific kind of representational knowledge of the mind and the appropriate age level and matched control groups have yet to be conducted.

What might be the source of this relatively spared capacity for mentalizing in Williams syndrome? One answer might be that this functional ability is directly related to the spared neural systems that have been identified in neuroimaging studies of Williams syndrome. In particular the medial temporal cortex, other parts of the limbic system, and orbito-frontal cortex have all been implicated in studies of normal adults engaged in mentalizing tasks, or in face processing (e.g., Baron-Cohen & Ring, 1994; Brothier & Ring, 1992; Perrett et al., 1990), and these same areas are known to be relatively spared on volumetric measures in the brains of subjects with Williams syndrome. Another answer might be that beginning in early infancy the difference between Williams syndrome and other retarded populations is that as babies and later as children they spend significantly longer looking at faces. This attentional difference is coupled with a strong interest in people, compared to objects, and a very outgoing personality. Over the course of development this unusual social attentional/personality profile leads to more opportunities to gain social information of the sort that is tapped in this mentalizing task. Future research should focus on confirming the links between these neurobiological and cognitive developmental interpretations of this spared cognitive capacity in Williams syndrome.

The adults with Prader-Willi syndrome performed quite poorly on this task, about as poorly as the subjects with autism who were tested by Baron-Cohen, Jolliffe, et al. (1997). The reason for poor performance on this mentalizing task may not be the same for these two distinct populations, especially because the subjects with Prader-Willi syndrome were mentally retarded whereas Baron-Cohen et al.'s autistic subjects were all of normal intelligence. The subjects with autism do poorly on this task because they have specific impairments in mentalizing ability. In contrast, the subjects with Prader-Willi syndrome do not seem to show specific problems in this general domain: We have some preliminary evidence to suggest that children with Prader-Willi syndrome pass theory of mind tasks at the same mental age levels as other matched mentally retarded children (Tager-Flusberg, Sullivan, Boshart, & Levine, 1996), in contrast to children with autism (Baron-Cohen et al., 1985; Baron-Cohen et al., 1993). Instead, there are probably general cognitive processing factors associated with low IQ, such as attention, response bias, and memory, that affect the Prader-Willi subjects' ability to perform well on this task. Although relatively little is known about the neuropathology associated with Prader-Willi syndrome,

a few studies have identified generalized reductions in cortical size, which may be a result of anomalies in cortical growth (Hayashi et al., 1992; Leonard et al., 1993; Reske-Nielsen & Lund, 1992). These pathological findings are consistent with the overall reduced cognitive capacities found in Prader-Willi syndrome and contrast with the more striking asynchronies in the size of different neural structures in Williams syndrome (Bellugi et al., 1994; Galaburda et al., 1994).

Despite relative strengths in interpreting mental state information from facial expressions, children and adults with Williams syndrome experience considerable difficulty in their social relationships, especially with peers (Gosch & Pankau, 1994). They seem unable to understand some of the complexities of social interactions and have limited understanding of friendship and other social concepts (Tager-Flusberg, Sullivan, Boshart, Guttman, & Levine, 1996). This suggests that the domain of social cognition, broadly defined, may not be as spared in Williams syndrome as one might predict from their performance on tasks tapping mentalizing ability. It is unlikely, therefore, that mentalizing and other aspects of social cognition form a unitary cognitive domain. We suggest that further research on children and adults with Williams syndrome may help to elucidate the definition and boundaries of this domain, and we can provide important clues to its cognitive architecture at both structural and functional levels.

METHODS

Subjects

Three groups of subjects participated in this study: adults with Williams syndrome, adults with Prader-Willi syndrome, and normal adults.

Adults with Williams Syndrome

Thirteen adults with Williams syndrome participated in this study, eight men and five women, recruited from the biennial Family Convention of the Williams Syndrome Association. Subjects' ages ranged from 17;11 to 37;0, with a mean age (and standard deviation) of 27;3 (5;7).

Adults with Prader-Willi Syndrome

Thirteen adults with Prader-Willi syndrome participated in this study, five men and eight women, recruited from a local residential center for adults with Prader-Willi syndrome. Their ages ranged from 22;11 to 42;4, with a mean age (and standard deviation) of 31;0 (6;5).

Normal Adults

Twenty-five normal adults participated in this study, nine men and sixteen women, recruited from an introductory psychology course. Although these adults were drawn from a college sample, we should note that the particular university from which they were drawn serves a diverse urban population. Subjects received credit for their participation. Ages ranged from 18;1 to 60;11, with a mean age (and standard deviation) of 26;4 (11;7).

All the subjects were individually tested on a standardized measure of verbal ability and a standardized IQ test. The PPVT-R (Dunn & Dunn, 1981) was used to assess receptive vocabulary, and the Kaufman Brief Intelligence Test (KBIT, Kaufman & Kaufman, 1990) was used to assess IQ. This test provides a composite IQ score based on subtests tapping verbal and nonverbal matrices reasoning ability. The subjects in the clinical groups all fell in the mild or moderate range of mental retardation; the normal subjects were selected for IQs falling within one standard deviation of the population mean. Table 3 shows mean age, IQ, and PPVT-R mental ages for the three groups of subjects.

The three groups were well-matched on age ($F(2, 49) = 0.33, ns$), and the two clinical groups were also matched on IQ ($F(1, 24) = 0.13, ns$) and the PPVT-R mental age equivalent score ($F(1, 24) = 0.10, ns$).

Procedures

The subjects were individually tested on the Eyes task as developed by Baron-Cohen, Jolliffe, et al. (1997) for use with normal adults as well as high-functioning adults with autism or Asperger syndrome. The stimuli consisted of 25 black-and-white photographs of the eye region of faces, taken from larger photographs in popular magazines. Each pair of eyes was digitized and cropped such

Table 3. Means (Standard Deviations) of Subject Characteristics

	Group		
	<i>Normal</i>	<i>Williams Syndrome</i>	<i>Prader-Willi Syndrome</i>
Male/Female	9/16	8/5	5/8
Age	26;4 (11;7)	27;3 (5;7)	31;0 (6;5)
IQ composite score	101.0 (9.7)	62.8 (9.7)	56.7 (9.8)
PPVT-R mental age	24;5 (7;6)	12;10 (6;7)	9;6 (2;8)

that the remaining image was black and white and measured 2 by 5 in., showing the eyes from just above the eyebrows down to the bridge of the nose. Baron-Cohen, Jolliffe, et al. (1997a) asked four judges (two male, two female) to generate labels for each picture in open discussion. These labels were then tested with their semantic opposites as foils on eight independent judges (four male, four female) who were blind to the hypotheses of the study. These eight judges unanimously agreed on the target labels for all items.

The photographs were shown to the subjects in this study one at a time in random order, and the target mental state term and its semantic opposite were read to the subject in counterbalanced order, such that for half of the items, the first term read was the correct response and for the other half the second term read was the correct response. Two versions of the task were prepared with the order of correct/incorrect terms reversed on the second version. Half the subjects within each group received one version, half the other. The full set of target and foil terms used on this task is shown in the "Appendix." For some of the items, the vocabulary words were also simplified to make them more accessible to the Williams syndrome and Prader-Willi syndrome subjects (e.g., for "Reflective/Unreflective" "Thoughtful/Not thoughtful" was added). With these populations, the original labels were read and then the simplified version, such that all subjects heard the original target and foil terms.

Appendix

Target Terms and Foil Terms for the Eyes Task

Item No.	Target Term	Foil Term
1	Concerned	Unconcerned
2	Noticing you	Ignoring you
3	Attraction	Repulsion
4	Relaxed	Worried
5	Serious message	Playful message
6	Interested	Not interested
7	Friendly	Hostile (Not friendly)
8	Sad reflection	Happy reflection
9	Sad thought	Happy thought
10	Certain	Uncertain
11	Far away focus (Looking far away)	Near focus (Looking near)
12	Reflective (Thoughtful)	Unreflective (Not thoughtful)
13	Reflective (Thoughtful)	Unreflective (Not thoughtful)

Target Terms and Foil Terms for the Eyes Task (cont.)

Item No.	Target Term	Foil Term
14	Cautious about something over there	Relaxed about something over there
15	Noticing someone else	Noticing you
16	Calm	Anxious
17	Dominant	Submissive
18	Fantasizing (Imagining)	Noticing
19	Observing	Daydreaming
20	Desire for you	Desire for someone else
21	Noticing you	Ignoring you
22	Nervous about you	Interested in you
23	Flirtatious	Not interested
24	Sympathetic	Unsympathetic
25	Decisive	Indecisive

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REFERENCES

- Baron-Cohen, S. (1995). *Mindblindness: An essay on autism and theory of mind*. Cambridge, MA: MIT Press.
- Baron-Cohen, S., & Hammer, J. (1997). Parents of children with Asperger's syndrome: What is the cognitive phenotype? *Journal of Cognitive Neuroscience*, 9, 548-554.
- Baron-Cohen, S., Jolliffe, T., Mortimore, C., & Robertson, M. (1997). Another advanced test of theory of mind: Evidence from very high functioning adults with autism or Asperger syndrome. *Journal of Child Psychology and Psychiatry*, 38, 813-822.
- Baron-Cohen, S., Leslie, A. M., & Frith, U. (1985). Does the autistic child have a "theory of mind"? *Cognition*, 21, 37-46.
- Baron-Cohen, S., & Ring, H. (1994). A model of the mindreading system: Neuropsychological and neurobiological perspectives. In C. Lewis & P. Mitchell (Eds.), *Children's early understanding of mind* (pp. 183-210). Hillsdale, NJ: Erlbaum.
- Baron-Cohen, S., Tager-Flusberg, H., & Cohen, D. J. (1993). *Understanding other minds: Perspectives from autism*. Oxford: Oxford University Press.

- Baron-Cohen, S., Wheelwright, S., & Jolliffe, T. (1997). Reading the mind in the face: Is there a "language of the eyes"? *Visual Cognition*, 4, 311-331.
- Bauman, M. L., & Kemper, T. L. (1994). Neuroanatomic observations of the brain in autism. In M. L. Bauman & T. L. Kemper (Eds.), *The neurobiology of autism* (pp. 119-145). Baltimore: Johns Hopkins University Press.
- Bellugi, U., Bihrlé, A., Neville, H., Jernigan, T., & Doherty, S. (1992). Language, cognition, and brain organization in a neurodevelopmental disorder. In M. Gunnar & C. Nelson (Eds.), *Developmental behavioral neuroscience* (pp. 201-223). Hillsdale, N.J.: Erlbaum.
- Bellugi, U., Marks, S., Bihrlé, A., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams syndrome. In D. Bishop & K. Mogford (Eds.), *Language development in exceptional circumstances* (pp. 177-184). New York: Churchill Livingstone.
- Bellugi, U., Wang, P., & Jernigan, T. L. (1994). Higher cortical functions: Evidence from specific genetically based syndromes of disorder. In S. Broman & J. Grafman (Eds.), *Cognitive deficits in developmental disorders: Implications for brain function* (pp. 23-56). Hillsdale, N.J.: Erlbaum.
- Brothers, L., & Ring, B. (1992). A neuroethological framework for the representation of minds. *Journal of Cognitive Neuroscience*, 4, 107-118.
- Brothers, L., Ring, B., & Kling, A. (1990). Responses of neurons in the macaque amygdala to complex social stimuli. *Behavioral Brain Research*, 41, 199-213.
- Butler, M. G. (1990). Prader-Willi syndrome: Current understanding of cause and diagnosis. *American Journal of Medical Genetics*, 35, 319-332.
- Butler, M. G. (1994). Prader-Willi and Angelman syndromes: Examples of genetic imprinting in man. In D. K. Seth & S. Seth (Eds.), *Human genetics: New perspectives* (pp. 73-94). New Delhi, India: Omega Scientific Publishers.
- Courchesne, E., Saitoh, O., Yeung-Courchesne, R., Press, G. A., Lincoln, A. J., Haas, R. H., & Schreibman, L. (1994). Abnormalities of cerebellar vermal lobules VI and VII in patients with infantile autism: Identification of hypoplastic and hyperplastic subgroups by MR imaging. *American Journal of Roentgenology*, 162, 123-130.
- Dunn, L. M., & Dunn, L. M. (1981). *PPVT: Peabody Picture Vocabulary Test-Revised*. Circle Pines, MN: American Guidance Services.
- Dykens, E. (in press). Prader-Willi syndrome: Towards a behavioral phenotype. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders: Contributions to a new framework from the cognitive neurosciences*. Cambridge, MA: MIT Press.
- Eslinger, P., & Damasio, A. (1985). Severe disturbance of higher cognition after bilateral frontal lobe ablation: Patient E. V. R. *Neurology*, 35, 1731-1741.
- Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., Spallone, P., Dean Stock, A., Leppert, M., & Keating, M. T. (1993). Hemizyosity at the elastin locus in a developmental disorder: Williams syndrome. *Nature Genetics*, 5, 11-16.
- Frangiskakis, J. M., Ewart, A. K., Morris, C. A., Mervis, C. B., Bertrand, J., Robinson, B. F., Klein, B. P., Ensing, G. J., Everett, L. A., Green, E. D., Proschel, C., Gutowski, N., Noble, M., Atkinson, D. L., Odelberg, S. J., & Keating, M. T. (1996). LIM-kinase1 hemizyosity implicated in impaired visuospatial constructive cognition. *Cell*, 86, 59-69.
- Galaburda, A. M., Wang, P. P., Bellugi, U., & Rossen, M. (1994). Cytoarchitectonic anomalies in a genetically based disorder: Williams syndrome. *Cognitive Neuroscience and Neuropsychology, Neuroreport* 5, 753-757.
- Gosch, A., & Pankau, R. (1994). Social-emotional and behavioral adjustment in children with Williams-Beuren syndrome. *American Journal of Medical Genetics*, 52, 291-296.
- Happé, F. (1994). An advanced test of theory of mind: Understanding of story character's thought and feelings by able autistic, mentally handicapped and normal children and adults. *Journal of Autism and Developmental Disorders*, 24, 129-154.
- Hayashi, M., Itoh, M., Kabasawa, Y., Hayashi, H., Satoh, J., & Morimatsu, Y. (1992). A neuropathological study of a case of the Prader-Willi syndrome with an interstitial deletion of the proximal long arm of chromosome 15. *Brain Development*, 14, 58-62.
- Jernigan, T. L., & Bellugi, U. (1990). Anomalous brain morphology on magnetic resonance imaging in Williams and Down syndrome. *Archives of Neurology*, 47, 529-533.
- Jernigan, T. L., & Bellugi, U. (1994). Neuroanatomical distinctions between Williams and Down syndrome. In S. Broman & J. Grafman (Eds.), *Atypical cognitive deficits in developmental disorders: Implications for brain function* (pp. 57-66). Hillsdale, N.J.: Erlbaum.
- Jernigan, T. L., Bellugi, U., Sowell, E., Doherty, S., & Hesselink, J. R. (1993). Cerebral morphological distinctions between Williams and Down syndrome. *Archives of Neurology*, 50, 186-191.
- Kamiloff-Smith, A., Klima, E., Bellugi, U., Grant, J., & Baron-Cohen, S. (1995). Is there a social module? Language, face processing and theory of mind in individuals with Williams syndrome. *Journal of Cognitive Neuroscience*, 7, 196-208.
- Kaufman, A. S., & Kaufman, N. L. (1990). *KBIT: Kaufman Brief Intelligence Test*. Circle Pines, MN: American Guidance Services.
- Ledbetter, D. H., Riccardi, V. M., Youngblom, S. A., Strobel, R. J., Keenan, B. S., Crawford, J. D., & Louro, J. M. (1980). Deletion (15q) as a cause of the Prader-Willi syndrome (PWS). *American Journal of Human Genetics*, 32, 77A.
- Leonard, C. M., Williams, C. A., Nicholls, R. D., Agee, O. F., Voeller, K. K., Honeyman, J. C., & Staab, E. V. (1993). Angelman and Prader-Willi syndrome: A magnetic resonance imaging study of differences in cerebral structure. *American Journal of Medical Genetics*, 46, 26-33.
- Mervis, C. B., & Bertrand, J. (1997). Developmental relations between cognition and language: Evidence from Williams syndrome. In L. B. Adamson & M. A. Ronski (Eds.), *Research on communication and language disorders: Contributions to theories of language development*. Baltimore: Paul Brookes.
- Minshew, N. J., Sweeney, J. A., & Furman, J. M. (1995). Evidence for a primary neocortical systems of abnormality in autism. *Society of Neuroscience Abstracts*, 21, 735.
- Morris, C. A., Ewart, A. K., Sternes, K., Spallone, P., Stock, A. D., Leppert, M., & Keating, M. T. (1994). Williams syndrome: Elastin gene deletions. *American Journal of Human Genetics*, 55 Supplement, A89.
- Nicholls, R. D., Knoll, J., Butler, M. G., Karam, S., & Lalande, M. (1989). Genetic imprinting suggested by maternal heterodisomy in non-deletion Prader-Willi syndrome. *Nature*, 342, 281-285.
- Numenmaa, T. (1964). *The language of the face. Jyväskylä studies in education, psychology and social research*. Jyväskylä, Finland: University of Jyväskylä.
- Perrett, D., Harries, M., Mistlin, A., Hietanen, J., Benson, P., Bevan, R., Thomas, S., Oram, M., Ortega, J., & Brierley, K. (1990). Social signals analyzed at the single cell level:

- Someone is looking at me, something touched me, something moved! *International Journal of Comparative Psychology*, 4, 25-55.
- Price, J. L., Russchen, F. T., & Amaral, D. G. (1987). The amygdaloid complex. In L. W. Swanson, A. Bjorklund, & T. Hokfelt (Eds.), *Handbook of chemical neuroanatomy*, vol 5, (pp. 297-388). New York: Elsevier.
- Reske-Nielsen, E., & Lund, E. (1992). Prader-Willi syndrome and central nervous system calcifications: Chance or fundamentally related findings? *Clinical Neuropathology*, 11, 6-10.
- Tager-Flusberg, H., & Sullivan, K. (1996). *Theory of mind abilities in young children with Williams syndrome*. Cognitive Neuroscience Society, April 1996, San Francisco, CA.
- Tager-Flusberg, H., Sullivan, K., Boshart, J., Guttman, J., & Levine, K. (1996). *Social cognitive abilities in children and adolescents with Williams syndrome*. Williams Syndrome Association, National Conference, July 1996, Philadelphia, PA.
- Tager-Flusberg, H., Sullivan, K., Boshart, J., & Levine, K. (1996). *Social relationships and social understanding in children with Prader-Willi Syndrome*. Northeast Regional Conference of the Prader-Willi Association, April 1996, Albany, NY.
- Thompson, T., Butler, M., MacLean, W., Joseph, B., & Delaney, D. (in press). Cognition, behavior, neurochemistry and genetics in Prader-Willi syndrome. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders: Contributions to a new framework from the cognitive neurosciences*. Cambridge, MA: MIT Press.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioral phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, 13, 232-244.
- Udwin, O., Yule, W., & Martin, N. (1987). Cognitive abilities and behavioral characteristics of children with ideopathic infantile hypercalcaemia. *Journal of Child Psychology and Psychiatry*, 28, 297-309.