



ELSEVIER

Available online at www.sciencedirect.com

SCIENCE @ DIRECT®

Research in Developmental Disabilities 27 (2006) 243–253

Research
in
Developmental
Disabilities

Do children with Williams syndrome fail to process visual configural information?

Christine Deruelle^{a,*}, Cécilie Rondan^{a,c},
Josette Mancini^b, Marie-Odile Livet^b

^a *Mediterranean Institute of Cognitive Neurosciences, CNRS, 31 chemin Joseph Aiguier, 13402 Marseille Cedex 20, France*

^b *Department of Pediatric Neurology, University Hospital La Timone, France*

^c *Laboratory of Psychology and Neurocognition, CNRS, UMR 5105, Grenoble, France*

Received 30 November 2004; received in revised form 18 February 2005; accepted 4 March 2005

Abstract

Configural visual abilities in thirteen children with Williams syndrome (WS) compared to 13 children matched on mental age and 13 children matched on chronological age. Configural abilities were tested through four tasks (1) Silhouette (2) Fragmented (3) Mooney and (4) overlapping figures. In the first three tasks, it was necessary to take into account the global information, as the identification of the figures could not be established through a local analysis. In the fourth task, the global configuration of the display had to be ignored. Configural skills seem appropriate in the WS population. A possible dissociation between perceptual and visuo-constructive configural competences is discussed.

© 2005 Elsevier Ltd. All rights reserved.

Keywords: Williams syndrome; Configural processing; Visual closure

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a microdeletion on one of the chromosome 7, comprising the area around the gene for elastin (Frangiskakis et al., 1996). Children with this syndrome often suffer from cardiovascular disease, musculoskeletal and renal abnormalities, hyperacusis, and facial dysmorphism (e.g., Laskari, Smith, & Graham, 1999). The syndrome is characterized by its

* Corresponding author. Fax: +33 4 91 77 49 69.

E-mail address: deruelle@incm.cnrs-mrs.fr (C. Deruelle).

distinctive and uneven cognitive profile (e.g., Jarrold, Baddeley, & Hewes, 1998). Individuals with WS show marked dissociations between language abilities that are relatively preserved in comparison to visuo-spatial skills that are particularly impaired. However, disparities are also observed within the visuo-spatial domain. Whereas face processing skills can be chronologically age-appropriate in this clinical pathology, studies have underlined particular weakness in visual tasks such as drawings and the block design task of the Wechsler scales (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000). Based on these observations, a generalized deficit of the dorsal visual stream has been hypothesized in the WS population (Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002). The anatomo-functional distinction between the ventral stream carrying information on object and facial identity and the dorsal stream conveying spatial relationship information seems to account quite well for the impaired spatial cognition and unimpaired face recognition observed in the WS pathology. However, some but not all aspects of face processing are preserved in children with WS (Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999). It thus appears at this point necessary to further precise the nature of visuo-spatial impairments in the WS individuals.

As no correlation has been found between the presence of a visual deficit such as strabismus, visual acuity loss or amblyopia and the severity of the visuospatial problem (Atkinson et al., 2001), authors have attempted to explain the visuo-spatial impairments by invoking an abnormal processing style. This processing style has been presumed to rely on local rather than on global information, contrary to typically developing children, who process global information in priority (e.g., Bellugi et al., 2000). Birhle, Bellugi, Delis, & Marks (1989) asked WS subjects to copy hierarchical forms composed of local elements arranged to form a global shape (for instance a capital D made of small Ss). They observed that WS individuals were more accurate in reproducing the local than the global aspects of the pattern whereas the control population (i.e., Down children) presented the opposite bias. Bertrand, Mervis, & Eisenberg (1997) reached the same conclusion. They tested the capacity of WS children in copying drawings of an elephant, a flower and a house. Again, results showed that WS accurately drew the details but not the global shape. Similarly, in the Block Design Task, WS had problems retrieving the overall global configuration of the cubes (Wang, Doherty, Rourke, & Bellugi, 1995).

Local processing has also been noticed in face discrimination tasks tapping configural analysis. Deruelle et al. (1999) showed that WS children were less efficient than their control-matched in processing the configural aspects of faces. Moreover, WS children presented a reduced effect of face inversion, which is an indication of local face processing. These data were in line with those obtained by Karmiloff-Smith (1997) in the same type of tasks. Taken together these diverse findings suggest that WS children exhibit difficulties in putting parts into a whole and in processing information about the configuration and the contour of shapes and faces. However, it is noteworthy that apart from the face tasks, most of the data suggesting configural impairments in WS individuals were collected through visuo-constructive tasks.

The main aim of this study was to further explore configural abilities in children with WS when considering the visuo-perceptual domain. The relative importance of configural and local visual processing was therefore examined through various experiments,

including three configural grouping and a disembedding tasks. The performances of the children with WS were compared to two groups of typically developing children, a group matched on verbal mental age and a group matched on chronological age.

In the three configural tasks, the visual target can be matched to its probe only if a holistic percept can be achieved from fragmentary information. In these tasks, featural information are either non available or non relevant. In the disembedding task, visual targets can be detected only by ignoring larger configurations which are task-irrelevant but which are automatically processed by typically developing children.

1. Configural grouping tasks

Each task in this part of the study needed the combination of fragmented information to form larger configurations. In other words, the perceiver must combine a number of individually meaningless parts to form a structured whole. If the hypothesis that configural processing is deficient in WS children is verified, they should then exhibit more difficulties than control participants in these tasks.

1.1. Silhouette identification task

This task was an attempt to verify whether WS participants were able to use global information in the absence of local internal features. Silhouette figures were obtained by darkening the internal features of line drawings so that the whole shape was black. The only information available on the silhouettes was information on the contour of the object. If WS children represent objects in terms of features in priority, then removal of details in a silhouette drawing should impair their ability to identify the pictures. They should exhibit poorer performance than controls in this task.

1.1.1. Method

1.1.1.1. Participants. A total of 39 children participated in the four experiments. Participants were divided into three groups, a group of 13 participants with WS (5 boys and 8 girls), 13 children matched individually with a child with WS on gender and mental age (MA), and 13 children matched on gender and chronological age (CA) using a 3 months gap. The children with WS were mostly recruited via the help of Regional Williams Syndrome Associations. Pediatric geneticists established WS diagnoses. The children fulfilled the criterion set out by Preus (1984) for a diagnosis of Williams syndrome. All but two WS participants received genetic testing and those tested were positive on the FISH test for elastin deletion on a copy of chromosome 7. Their ages ranged from 5 years 11 months to 17 years 3 months ($M = 10$ years 5 months, $S.D. = 3$ years 7 months). At the time of testing, all the children attended schools or specialized centers for the educable mentally retarded. Mental age, inferred from IQ measures (WISC-R (Wechsler, 1996) or WPPSI (Wechsler, 1995) according to the subject's age), ranged from 4 to 8 years 3 months ($M = 5$ years 4 months, $S.D. = 1$ year 3 months). IQ scores were within the range given for WS individuals as reported by several recent studies (e.g., Jarrold et al., 1998).

In the MA-matched group, children were aged from 4 years 9 months to 6 years ($M = 5$ years 5 months, $S.D. = 1$ year 3 months) and in the CA-matched group, the children were aged from 5 years 8 months to 17 years ($M = 10$ years 7 months, $S.D. = 3$ years 7 months).

1.1.1.2. Stimuli and procedure. The stimulus set was composed of 26 colored drawings of animate and inanimate objects (18 pictures of animals, 4 pictures of cars and 4 pictures of flowers) and their corresponding silhouettes.

Participants underwent 36 test trials and 2 training trials. During a trial, three stimuli appeared simultaneously on the screen, a target at the top and two probe stimuli side by side at the bottom of the screen. The target was one of the 26 colored drawings. The positive probe was its silhouette and the negative probe was another object belonging to the same category as the target. Negative probes matched the target on its size and general shape. Two conditions composed this experiment, one condition (same) in which the target and its probes were displayed with the same orientation ($N = 24$ trials) and another condition (different) in which the probe stimuli were oriented 45° compared to the target ($N = 12$). This latter condition was chosen to render the task more difficult.

Participants sat in front of a portable computer (Macintosh). Distance between the subject and the screen was set at approximately 49 cm. Participants were tested individually at their home or in a laboratory office at the CNRS.

They were asked to find which probe corresponded to the target. The position (left or right) of the positive probe was counterbalanced across trials. Participants had to press the 'a' button if they chose the silhouette on the left side of the screen or the 'p' button if they chose the silhouette on the right side of the screen. Different colored patches identified keyboard buttons.

1.1.2. Results and discussion

A 2×3 ANOVA with condition (same/different) as a within factor and group (WS/MA-matched/CA-matched) as a between factor was conducted on the percentage of errors. The group effect was significant ($F(2-36) = 5$, $p < .05$), revealing a significant difference between the MA-matched and the CA-matched group ($F(1-24) = 10$, $p < .01$), but not between the WS and the MA-matched nor between the WS and the CA-matched (all $ps > .05$). The condition factor was significant ($F(1,36) = 15.7$, $p < .001$), showing better performance in the Same ($M = 6.6$, $S.D. = 5.5$) than in the different ($M = 13.3$, $S.D. = 10$) condition. The interaction between groups and conditions was not significant ($F(2,36) = 3$, $p > .05$). Mental rotation of the target indeed made the task harder for both WS and control participants.

In order to check if age improved performance a Spearman rank correlation was computed between the chronological age of the WS subjects and their number of errors. The correlation between these two factors was not significant ($ps > .05$).

1.2. Fragmented objects task

If children with WS exhibit a local bias and have difficulties in processing configurations, they should be less efficient than controls in identifying objects that are contour-deleted.

1.2.1. Method

The same participants as in the Silhouette identification task participated in this task.

The stimuli were selected from the Fragmented Picture Test (Snodgrass & Vandewart, 1980). Pictures of 48 line-drawn objects, familiar to all children (see the list of the 48 objects selected for this task in the footnote section¹), were digitized and degraded by deleting approximately 50% of their contour.

Sixteen displays were constructed with these 48 objects. Each display contained a target, which was a complete line-drawn object presented at the top the screen, and four fragmented probe stimuli presented horizontally at the bottom of the screen. The probe stimuli consisted of the fragmented version of the target stimuli and three distractors. Distractors were chosen to be structurally or semantically close to the target. The fragmented version of each target was used as a distractor in another display. Each object (fragmented or not) subtended $1^\circ \times 2^\circ$ of visual angle when viewed at 49 cm from the screen.

Each participant was presented with the 16 displays. They were asked to recognize the target among the four fragmented objects situated at the bottom of the portable screen. They were to point with their finger to the correct match. A keyboard button was dedicated to each of the four fragmented objects ('a' for the most left, 'r' for intermediate left, 'u' for the intermediate right and 'p' for the most right). Once the child had made his choice, the experimenter pressed the keyboard button corresponding to his response. The order of presentation was counterbalanced across subjects.

The remaining procedure was identical to the previous task.

1.2.2. Results and discussion

An ANOVA with group (WS/MA-matched/CA-matched) as a between factor was performed on the number of errors. The group effect was significant ($F(2-36) = 6$, $p < .05$), showing that performance of CA-matched children ($M = 96\%$ of correct answers) was better than that of both WS ($M = 80\%$ of correct answers; $F(1-24) = 10$, $p < .01$) and MA-matched ($M = 87\%$ of correct answers; $F(1-24) = 7.5$, $p < .01$). In contrast, WS did not differ from MA-matched children ($p > .05$). Moreover, inspection of the responses by WS revealed that they made the same type of errors as their MA-matched controls. The most frequent errors were the confusion between the axe and the key (16% of total errors in the WS group and 7% in the MA-matched group), between the brush and the pencil (14% of errors in the WS group and 12% in the MA-matched group) and between the chair and the rocking chair (11% of errors in the WS group and 17% in the MA-matched group). It should be underlined that performance of CA-matched was possibly at ceiling in this task and may be difficult to interpret.

The number of errors did not vary with mental nor chronological age in the WS population (Spearman rank correlation test, $ps > .05$).

¹ List of the 48 objects used in Experiment 1: brush, pen, glass, chair, guitar, violin, apple, lemon, key, pencil, watch, peach, axe, saw, fork, ruler, screwdriver, hat, pear, bell, balloon, bed, bus, airplane, helicopter, rocking chair, tomato, pliers, car, pumpkin, table, banana, knife, harp, cradle, peach, barrel, sailboat, paintbrush, anchor, rolling pin, candle, clock, toothbrush, belt, clock, comb, spoon.



Fig. 1. Example of display used in the Mooney face task.

1.3. Mooney face task

This experiment used high contrasted pictures of faces containing only black (shadow) and white (highlight) gratings (see Fig. 1). As in the previous tasks, one must achieve a configural percept from fragmentary information in order to match the photographs correctly (Wasserstein, Zappulla, Rosen, Gerstman, & Rock, 1987). The Mooney face tasks may not be tackled by using only individual features perception. If WS children focus on local information and do not integrate featural information into a coherent whole, then they should be impaired in this task.

Previous researchers have used similar stimuli and have reported intact visual closure abilities in WS persons (Jones, Hickok, & Lai, 1998; Wang et al., 1995). However, the typical Mooney face task (Mooney, 1957) requires categorization processes that may lead to different results than matching processes. Moreover, the earlier Mooney task studies compared the performance of children with WS with that of children with Down syndrome (Jones et al., 1998; Wang et al., 1995), but not with typically developing children, as it is the case in the current series of experiments. Finally, we wished to compare the performance obtained on the same group of participants in various tasks pinpointing configural analysis.

1.3.1. Method

The same subjects as in the two previous tasks participated in this experiment.

Stimuli were constructed from 20 black and white photographs of 10 faces taken from two viewpoints. Each of these 20 photographs was high contrasted using Adobe Photoshop software in order to leave only patches of light and shadow and no outer contour. Each Mooney face subtended $6^\circ \times 6^\circ$ of visual angle when viewed at 49 cm.

This experiment was composed of two conditions: a ‘Structural’ and an ‘Identity’ condition. Three Mooney faces were simultaneously presented on the screen, one target at the top and two probe stimuli at the bottom of the screen. In the ‘Structural’ condition, the positive probe stimulus was the same Mooney as the target and the negative probe was the Mooney face of another person. In the ‘Identity’ condition, the positive probe was the Mooney face of the same person as the target but presented from a different viewpoint and the negative probe was a Mooney face of another person. Position, left or right, of the positive probe was counterbalanced across trials. Positive probe in one trial was systematically the negative probe in another trial. After a few trials of training ($N = 5$), each participant firstly underwent 10 trials in the ‘Structural’ condition and then 10 trials in the ‘Identity’ condition.

The remaining procedure was identical to that of the previous experiments.

1.3.2. Results

A 2×3 ANOVA with condition (structural/identity) as a within factor and group (WS/MA/CA) as a between factor was conducted on the number of errors. The group factor was not significant ($F(2-36) = .05, p > .10$) indicating that WS subjects performed equally well as both MA-matched and CA-matched controls. The condition factor was significant ($F(1-36) = 73.7, p < .001$) revealing higher performance in the Structural ($M = 0.3, S.D. = 0.6$) than in the Identity ($M = 2.8, S.D. = 1.6$) conditions. The group by condition interaction was not significant ($p > .05$) and the condition effect was significant within the three groups of children (respectively, WS: $F(1-12) = 23.2, p < .001$); MA-matched: $F(1-12) = 23, p < .001$; CA-matched: $F(1-12) = 28, p < .001$).

In the WS population, performance did not depend on neither mental nor chronological age (Spearman rank correlation test, $ps > .05$).

Taken together, the results obtained in these numerous configural tasks showed that WS individuals performed as well as their MA-matched controls in all tasks, and as well as their CA-matched in all but one task, indicating that visual grouping abilities are mostly intact in this population. Moreover, both WS and control participants had most difficulties in the most complicated conditions (i.e., ‘Identity’ condition in the Mooney task and ‘Different’ condition in the Silhouette task), suggesting that WS participants performed the tasks in the same manner as their controls.

2. Overlapping figures

This task requires that participants ignore larger configurations in order to see targets at the local level. In contrast to the previous configural tasks, WS children should perform better than controls if they are less sensitive to configural information.

2.1. Method

This experiment consisted of two conditions: a Face and a Non-Face condition. In the Face condition, stimuli set comprised 10 line-drawn faces of children (5 male and 5 female faces). In the non-face condition, stimuli set comprised 24 line drawings of familiar objects

and 10 line drawings of animals. Sizes of line-drawn objects and faces were approximately the same. Each condition contained 10 displays that were made of one target in the center and two positive and two negative probes, each situated in one of the four corners. A target was constructed by juxtaposing two line drawings, positives probes were the two line drawings used in the target and the two negative probes were two other line drawings. In the Face condition, targets were made of two line drawings of faces and in the non-face condition; targets were made of line drawings of two animals ($N = 4$ trials) or of two objects ($N = 6$ trials). Negative probes belonged to the same category as the targets. Position of the positive probes was counterbalanced across displays. Each positive probe in one display was used as a negative probe in another display. Order of presentation was randomized for each participant. Half of the participants began with the face condition and the other half began with the non-face condition. Laser printed versions of the displays were used. Subjects were asked to find among the four drawings located in each corner of the sheet of paper the drawings constituting the target. Subjects could either trace a line between the target and his/her choice or just point to his/her choice and the experimenter traced the line for him/her.

Every correct response was scored 1 so the maximum score was 20 for each condition.

2.2. Results and discussion

Data from CA-matched subjects were not included in the analysis because of ceiling performance. A 2×2 ANOVA with condition (face/non-face) as a within factor and group (WS/MA-matched) as between factor were performed on the score. There was no effect of either group or condition (all $ps > .05$). Both WS individuals ($M = 90\%$ correct) and MA subjects ($M = 91\%$ correct) performed equally well at this task.

Potential differences in the two groups or the two conditions may have been masked by the rather simple task.

3. General discussion

Children with WS participated in a series of configural tasks in order to verify whether they possessed some or no configural processing capacities. The performance of the WS children is very similar to that of controls. These data may appear in sharp contrast with those showing a configural deficit in this population (Birhle et al., 1989).

So far, the visuo-spatial deficit in the WS population has mostly been documented in experiments relying on spatial components. The initial report that established a configural deficit in WS children concerned a drawing task. As earlier mentioned, Birhle et al. (1989) examined the performance of WS children when drawing and copying hierarchical stimuli. In this task, WS participants accurately drew the local features but their drawings did not include the global configuration of the form. Similar copying and spontaneous drawing deficits were reported in a longitudinal case study of a WS girl (Stiles, Sabbadini, Capirci, & Volterra, 2000). These reports underline that drawings by subjects with WS seem to lack the configural organization.

Additionally, WS children show a selective disability on items requiring the integration of component parts as in the test of visual-motor integration (VMI: Bellugi, Wang, &

Jernigan, 1994). Furthermore, several authors have found a severe impairment in WS children in the block design task (Rondan, Mancini, Livet, & Deruelle, 2003). This task requires participants to assemble three-dimensional cubes into a coherent whole in order to match a two-dimensional model. To succeed the task one must perceive the configuration of the pattern correctly and plan the movement before executing the action.

Based on these last reports, it appears that WS children show configural processing deficits. This is in opposition with the findings of the current study.

However, recent studies have pointed out that the visuo-spatial deficit typically described in WS population concerns visuo-constructive tasks, such as drawings and block design tasks, rather than visual tasks per se. Pani, Mervis, & Robinson (1999) demonstrated that WS individuals' performance was similar to that of control participants in a visual search task that involved grouping processes. More interesting for our purpose, Rondan et al. (2003) showed that though children with WS were severely comprised on the block design task, they performed comparably to age-matched controls on the perceptual component of this same task. Consistent with this, Farran, Jarrold, & Gathercole (2003) revealed that WS children performed as controls in a hierarchical pattern visual identification task but showed poorer performance in the drawing task.

These last data, along with the results of the current study, may indicate that perceptual abilities are preserved in the WS population, though their difficulties are of a more spatial configural nature.

However, this argument may be criticized by at least two findings. Why would visual configural abilities be preserved in the context of pattern processing, as in this study, but not in that of face processing (Deruelle et al., 1999)? Even more contradictory, the Mooney faces matching task which is a task that involved faces and configural processing, is performed correctly by WS individuals in the current study. It seems difficult to reconcile this finding with the configural face processing impairment observed in Deruelle et al. (1999).

One may argue that it is very unlikely that only configural face processing is impaired while configural object processing is not. Firstly, configural deficits were found in visuo-constructive tasks on geometrical patterns. Secondly, the Mooney task is more correlated to visual closure abilities than to face processing. Previous results from the literature on visual cognition suggest that performance on the Mooney tend to group with performance on other closure tasks, rather than with facial perceptual tasks (Wasserstein et al., 1987).

One possible explanation of the discrepancies in results could be that the level of task difficulty may vary between this study and the previous studies on faces. However, this hypothesis is not confirmed. Inspection of the overall level of performance shows that WS participants performed at 87.7% correct in average in the faces tasks of Deruelle et al. (1999) and at 81% correct in average in the current study.

Another explanation could be that configural processing is not unique and that some, but not all configural processes are preserved in WS persons. Configural processing such as the one tested in face tasks is possibly not the same as the one tested in visual closure abilities, as in the present report. This hypothesis is supported by neuropsychological findings. Recent studies have shown that holistic integration relies on the integrity of occipito-temporal cortex including the fusiform gyri (Humphreys, 1999). However, data on visual agnosia suggest that processes that integrate elements into contours are intact when these

brain regions are lesioned (Humphreys, 1999). This possibly means that holistic integration can be distinguished from the integration of elements into contours. WS persons could thus be impaired in holistic integration of facial features (as in Deruelle et al. (1999) and in Karmiloff-Smith, 1997) but not in integration of elements into contours (as in this present study). If verified, this functional distinction needs to be taken into account in the elaboration of remediation programs.

Acknowledgments

We are grateful to the French Regional Associations of Williams Syndrome (Associations du Sud Est and Rhone Alpes) and to the children and their parents for their participation in the study.

References

- Atkinson, J., Anker, S., Braddick, O., Nokes, L., Mason, A., & Braddick, F. (2001). Visual and visuospatial development in young children with Williams syndrome. *Developmental Medicine and Child Neurology*, *43*, 330–337.
- Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St. George, M. (2000). The neurocognitive profile of Williams syndrome: a complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*, *12*, 7–29.
- Bellugi, U., Wang, P. P., & Jernigan, T. L. (1994). Williams syndrome: An unusual neurophysiological profile. In S. Broman, & J. Grafman (Eds.), *Atypical cognitive deficits in developmental disorders: Implications for brain function* (pp. 23–56). Hillsdale, NJ: Erlbaum.
- Bertrand, J., Mervis, C. B., & Eisenberg, J. D. (1997). Drawing by children with Williams syndrome: A developmental perspective. *Developmental Neuropsychology*, *13*, 41–67.
- Birhle, J. M., Bellugi, U., Delis, D., & Marks, S. (1989). Seeing either the forest or the trees: Dissociation in visuospatial processing. *Brain and Cognition*, *11*, 37–49.
- Deruelle, C., Mancini, J., Livet, M. O., Cassé-Perrot, C., & de Schonen, S. (1999). Configural and local processing of faces in children with Williams syndrome. *Brain and Cognition*, *41*, 276–298.
- Farran, E. K., Jarrold, C., & Gathercole, S. E. (2003). Divided attention, selective attention and drawing: Processing preferences in Williams syndrome are dependent on the task administered. *Neuropsychologia*, *41*, 676–687.
- Frangiskakis, J. M., Ewart, A. K., Morris, C. A., Bertrand, J., Robinson, B. F., Klein, B. P., et al. (1996). Lim-Kinase-1 hemizyosity implicated in impaired visuospatial constructive cognition. *Cell*, *86*, 59–69.
- Humphreys, G. W. (1999). Interactive agnosia. In G. W. Humphreys (Ed.), *Case studies in the neuropsychology of vision* (1st ed., pp. 41–58). Hove: Psychology Press.
- Jarrold, C., Baddeley, A. D., & Hewes, A. K. (1998). Verbal and nonverbal abilities in Williams syndrome phenotype: evidence for diverging development trajectories. *Journal of Child Psychology and Psychiatry*, *39*, 511–523.
- Jones, W., Hickok, G., & Lai, Z. (1998). Does face processing rely on intact visual spatial abilities? Evidence from Williams syndrome. *Cognitive Neuroscience Society Abstract Program*, *80*, 67.
- Karmiloff-Smith, A. (1997). Crucial differences between developmental cognitive neuroscience and adult neuropsychology. *Developmental Neuropsychology*, *13*, 513–524.
- Laskari, A., Smith, A., & Graham, J. (1999). Williams-Beuren syndrome: an update and review for the primary physician. *Clinical Pediatrics*, *38*, 189–208.
- Mooney, C. M. (1957). Age in the development of closure ability in children. *Canadian Journal of Psychology*, *11*, 216–226.
- Pani, J. R., Mervis, C. B., & Robinson, B. F. (1999). Global organization by individuals with Williams syndrome. *Psychological Science*, *10*, 453–458.

- Paul, B. M., Stiles, J., Passarotti, A., Bavar, N., & Bellugi, U. (2002). Face and place processing in Williams syndrome: evidence for a dorsal-ventral dissociation. *NeuroReport*, *13*, 1115–1119.
- Preus, M. (1984). The Williams syndrome: Objective definition and diagnosis. *Clinical Genetics*, *25*, 422–428.
- Rondan, C., Mancini, J., Livet, M. O., & Deruelle, C. (2003). Perceptual and visuo constructive performance in children with Williams syndrome. *Cognition, Brain, Behavior*, *7*, 149–157.
- Snodgrass, J. G., & Vandewart, M. (1980). A standardized set of 260 pictures: Norms for name agreement, image agreement, familiarity, and visual complexity. *Journal of Experimental Psychology: Human Learning*, *6*, 174–215.
- Stiles, J., Sabbadini, L., Capirci, O., & Volterra, V. (2000). Drawing abilities in Williams syndrome: A case study. *Developmental Neuropsychology*, *18*, 213–235.
- 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.
- Wasserstein, J., Zappulla, R., Rosen, J., Gerstman, L., & Rock, D. (1987). In search of closure: Subjective contour illusions, Gestalt completion tests, and implications. *Brain and Cognition*, *6*, 1–14.
- Wechsler, D., 1995. *Manual for the intelligence scale for preschool and primary children—revised*. The Psychological Corporation.
- Wechsler, D., 1996. *Manual for intelligence scale for children* (3rd ed). The Psychological Corporation.