Exp Brain Res (2004) 157: 137–146
DOI 10.1007/s00221-004-1834-0

RESEARCH ARTICLES

Maria-Alexandra Georgopoulos · Apostolos P. Georgopoulos · Nicole Kuz · Barbara Landau

Figure copying in Williams syndrome and normal subjects

Received: 6 August 2003 / Accepted: 29 December 2003 / Published online: 17 February 2004

Abstract We evaluated the copying abilities of ten subjects with Williams syndrome (WS; age 6–14 years) and ten normally developing children (age 3–6 years) matched for mental age using the matrices component of the Kaufman Brief Intelligence Test (mKBIT). Each subject copied six figures, including line drawings of closed and open geometrical shapes (alone and in combination), crossed lines, and geometrical shapes made of distinct small, filled circles. Qualitatively, subjects of both groups made comparable copies, although several subjects with WS drew a continuous line when copying figures composed of distinct circles. Quantitatively, the goodness of the copies was assessed by three human observers who rated on an analog scale the similarity of each copy to its visual template. Ratings were converted to a scale from zero (completely different) to 100 (the same) for statistical analyses. We found the following. First, the overall goodness of copies of the templates was very similar between the WS and control groups (WS: mean=46.7, range=0.89–95.4; control: mean=54.5, range=0.89–98.2). Second, there were systematic differences in the goodness of copies between the two groups, depending on the features of the figures. Specifically, the goodness of copies of control subjects was almost the same as that of WS subjects for simple line figures, but was consistently better for composite line figures, and even better for figures in which the shape was made of small, filled circles. Third, there was a significant relation between the goodness of copies (dependent variable) and mental age (mKBIT, independent variable) in both groups, although it was stronger and more highly statistically significant in the control than the WS group. These findings indicate that the principles guiding copying are similar in the two groups and suggest that WS is a case of developmental rather than deviance disorder.

Keywords Williams syndrome · Copying · Mental age

M.-A. Georgopoulos
Brain Sciences Center, Minneapolis Veterans Affairs Medical Center,
Minneapolis, MN, USA

M.-A. Georgopoulos
Department of Radiology, University of Minnesota Medical School,
Minneapolis, MN, USA

A. P. Georgopoulos
Departments of Neuroscience, Neurology and Psychiatry,
University of Minnesota Medical School,
Minneapolis, MN, USA

A. P. Georgopoulos (✉)
Brain Sciences Center (11B), VAMC,
One Veterans Drive,
Minneapolis, MN 55417, USA
e-mail: omega@umn.edu
Tel.: +1-612-7252282
Fax: +1-612-7252291

N. Kuz
Department of Psychology, University of Delaware,
Newark, DE, USA

B. Landau
Department of Cognitive Science, Johns Hopkins University,
Baltimore, MD, USA

Introduction

A cardinal feature of WS is disturbed copying (see Bellugi et al. 2000 for a review). Copies of figures are usually inaccurate reproductions of the template, and the severity of the disturbance varies among subjects. The nature of this dysfunction is unknown. Given that copying figures is a complex visuomotor skill, it could be affected by various factors and at different processing stages, from the beginning of the sensory processing of the visual stimulus to the motor drawing. For example, bad copies could be the result of abnormal visual perception, visual agnosia, motor deficits, involuntary movements, apraxia, or any combination thereof. By definition, abnormal copying is the main feature of constructional apraxia, which is a disorder of the translation of visuo-spatial input to a congruent spatio-motor output (Kleist 1934; Gainotti

1985). Subjects suffering from this disorder typically have intact visual and motor function but they have difficulty in copying figures. This deficit can be also observed when drawing figures from memory. Remarkably, copying from vision or drawing from memory can be differentially affected: single cases have been described of patients who copied well from vision but not from memory (Grossi et al. 1986), and vice versa (Servos and Goodale 1995). Finally, disturbed copying can also be dissociated from visual recognition of the figure; for example, a patient with visual agnosia could copy well (Behrmann et al. 1992), whereas another patient with intact visual recognition has copied abnormally (Cipolotti and Denes 1989). With respect to WS, the extent of the spatial impairment is currently unknown. However, several spatial capacities appear to be unimpaired, including visual gnosis (Hoffman and Landau 2000), perception of biological motion (Jordan et al. 2002), and many aspects of the use of spatial terms which presumably reflect conceptual understanding of spatial relations (Landau and Zukowski 2003). Therefore, the disturbance of copying seems to be a primary defect. In this respect, WS resembles the syndrome of developmental constructional apraxia (De Ajuriaguerra and Stambak 1969).

Most of the evaluation of the copying function in WS has relied on qualitative assessments (see, e.g., Bertrand et al. 1997; Bellugi et al. 2000; Mervis and Klein-Tasman 2000). In contrast, here we present a detailed formal method for evaluating the drawings which has not been done to date, to our knowledge. Specifically we employed a psychophysical assessment of the "goodness of copy" (with respect to the template) by human adult raters who also assessed the similarity of corresponding copies of mental-age-matched WS and control subjects. In this way we were able to assess not only the similarities between drawings but also their dependence on mental age. This psychophysical approach was necessary since, to our knowledge, there is no rigorous algorithmic procedure to evaluate the similarity between complex figures, unlike the case for simple shapes (Dryden and Mardia 1998).

Materials and methods

Subjects

Ten subjects (six females and four males) with WS syndrome and ten control subjects (seven females and three males) volunteered to participate in these studies. The study protocol was approved by the Institutional Review Board of the University of Delaware and informed consent was obtained from guardians prior to participation in the experiments. The subjects' informed consent was obtained according to the Declaration of Helsinki.

Kaufman Brief Intelligence Test (KBIT; Kaufman and Kaufman 1990)

This test was administered in accord with standardized instructions. The test has two components, a Verbal and a Matrices subtest. The Verbal test (vKBIT) comprises two subtests, in ascending difficulty. The first, simpler subtest requires the subject to name a set of objects presented as black and white drawings, whereas the second, more difficult subtest requires reading. Only the first subtest was administered in this study, for most subjects in both study groups did not read. Therefore, the "vKBIT" term is used below to refer to this simpler subtest.

The Matrices test (mKBIT) is a non-verbal test, in that it requires non-linguistic judgments of similarity and difference. It is not heavily loaded with spatial items, and hence does not unfairly penalize the WS children for their spatial deficit. The mKBIT items include simple problem-solving items, such as picking which picture "goes with" another, where the basis for matching is functional or thematic similarity (e.g. a hat goes on a head, a shoe goes on a foot, etc.). The problems become harder as one moves up the scale, and include pattern completion (e.g. selecting a pattern of dots that would complete a matrix of other dot patterns).

The subjects whose drawings were analyzed in this study were part of a larger group of WS and control subjects who were being tested on various tasks at that time. All subjects were matched on the basis of KBIT at the time they entered the study. The matching was done solely on the basis of KBIT score, without consulting the drawings.

Figure drawing

Templates

Each subject copied six figures (Fig. 1) which included line drawings of closed and open geometrical shapes (alone and in combination), crossed lines, and geometrical shapes made of small, filled circles. The figures were part of the Developmental Test of Visual-Motor Integration (Beery 1997).

Drawings

Subjects were shown each model on the upper half of an 81/2×11″ piece of paper, and were asked to copy the figure in the lower half of the paper. After they finished each drawing, they were also asked to describe the model by responding to the question, "What is this?" These data are not discussed in this paper; however, the children in both groups were comparably accurate in their descriptions of the models.

Rating the copies

Raters

Three adult subjects volunteered to rate the similarity of a drawing to its template, as described below. None of the raters were part of the research team. The protocol was approved by the appropriate

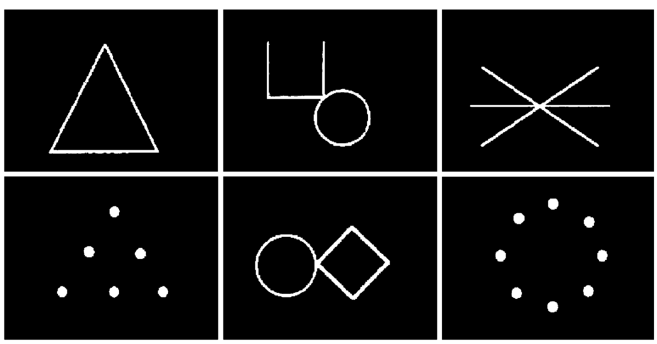

Fig. 1 The six figures presented as templates for copying

Institutional Review Boards and informed consent was obtained. The raters' informed consent was obtained according to the Declaration of Helsinki.

Procedure

The original templates and drawings by the subjects were displayed on a flat computer monitor as white on black background (see Fig. 2 for an example). Subjects rated the similarity of (a) drawings made by WS subjects to the corresponding template (group 1); (b) drawings made by control subjects to the corresponding template (group 2); and (c) drawings made by WS subjects to the corresponding drawings made by control subjects matched with respect to mKBIT (group 3: CWS). There were two arrangements for each template-drawing or drawing-drawing pair, left-right and right-left. This resulted in 10 matched subject pairs × 6 figures × 2 spatial arrangements × 3 groups = 360 displays which were shown to each rater in a completely randomized design. Randomization was done separately for each rater. All images were saved in Microsoft PowerPoint files and were presented by running this program.

Raters faced the computer monitor from a distance of ~76 cm at eye level. They were instructed to look at a pair of figures and rate as accurately as possible their similarity in an analog scale by placing a central vertical bar along a horizontal line the left and right ends of which were labeled Different and Same, respectively (Fig. 2). The image subtended ~9 deg and ~5 deg of visual angle in the horizontal and vertical dimension, respectively; the rating line also subtended ~9 deg of visual angle. There were no time restrictions placed on raters' reaction times. The images presented and the horizontal screen coordinate of the vertical bar after rating were saved and used for data analyses.

Data analysis

The similarity measure of pairs of figures was the horizontal screen coordinate of the rating bar. This was re-expressed on a scale from 0 (most different) to 100 (most similar) and averaged across raters and left-right arrangements. Thus 180 measurements were analyzed, namely 3 groups × 10 subjects/group × 6 figures/subject. Standard statistical methods were used to analyze the data (Snedecor and Cochran 1989). Probability levels given for the *t*-test are under the assumption of equal group variances, unless stated otherwise. The equality of variances assumption was assessed using Levene's test, and the *p* value of the *t*-test adjusted accordingly. All statistical analyses, including comparisons of means, analysis of variance, and regression analyses, were carried out using the SPSS (SPSS for Windows, version 10.1.0, SPSS Inc., Chicago, IL, 2000) and/or the BMDP/Dynamic (BMDP Statistical Software, Inc., Los Angeles, CA, 1992) statistical packages.

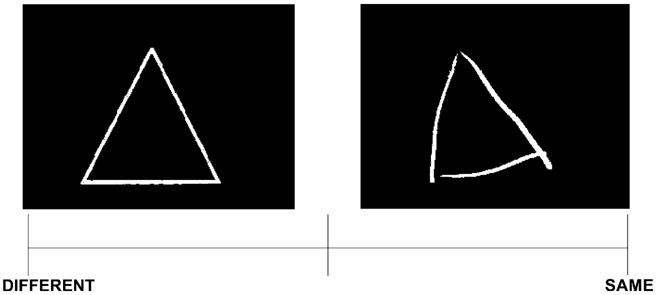

Fig. 2 Example of the display used for rating

Results

Analysis of subject characteristics

The WS and control groups were matched for mental age. Because WS individuals are generally mildly to moderately retarded, they were not matched for chronological age, and were older than their controls, as shown below.

Age and gender

The chronological age of subjects in the WS group was significantly greater (by 61.6 months) than that of subjects in the control group ($p<10^{-5}$, *t*-test, two-tailed, assuming unequal variances; $p=0.031$, Levene's test for equality of variances). The average chronological age of subjects in the WS group was 119.9±8.8 months (mean ± SEM, $N=10$; range=94 months); the ages of females (109±9.3, $N=6$) and males (136.25±14.6, $N=4$) did not differ significantly ($p=0.135$, *t*-test). The average age of subjects in the control group was 58.3±3.9 months (range=33 months); there was no significant difference between the ages of females (56.6±4.8, $N=7$) and males (62.3±7.5, $N=3$).

Mental age: vKBIT

The vKBIT score did not differ significantly between the WS and control groups [$p=0.498$, *t*-test; WS group (mean ± SEM): 29.35±1.95, control group: 27.1±2.6]. In addition, this score did not differ significantly between females and males in the WS group ($p=0.41$, *t*-test) or the control group ($p=0.17$).

There was a significant positive correlation between the vKBIT score and the chronological age in the control subjects ($r=0.767$, $p=0.01$, $N=10$ subjects) but not in WS subjects ($r=0.214$, $p=0.553$).

Mental age: mKBIT

The mKBIT score did not differ significantly between the WS and control groups [$p=0.315$, *t*-test; WS group (mean ± SEM): 18.1±1.11, control group: 16.4±1.21]. In addition, this score did not differ significantly between females and males in the WS group ($p=0.17$, *t*-test) or the control group ($p=0.77$).

Matching on mKBIT score

Subjects were matched on the mKBIT score. The Pearson product-moment correlation coefficient between these scores in the two groups was 0.971 ($p=0.000003$, $N=10$ pairs). Although the average matrices KBIT scores did not differ significantly between the two groups as a whole, i.e. when considered as independent groups (see above), the

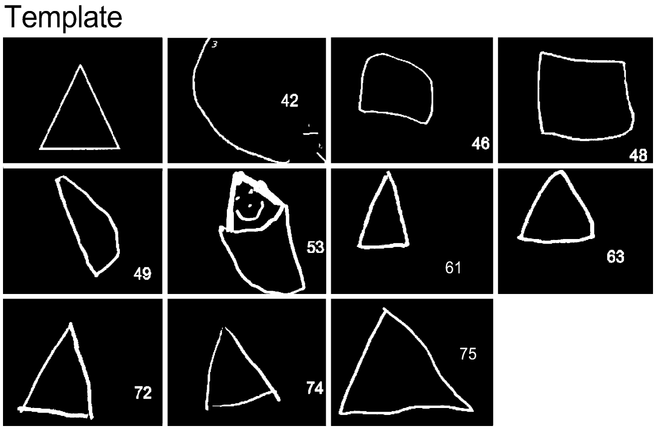

Fig. 3 Copies of the triangle by ten control subjects. *Numbers* are biological age in months

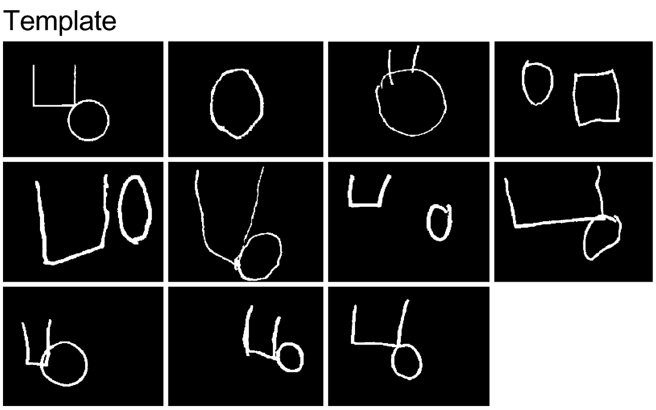

Fig. 4 Copies of another figure which is composed of two elementary shapes. Copies are arranged in the same sequence, with respect to subjects, as in Fig. 3

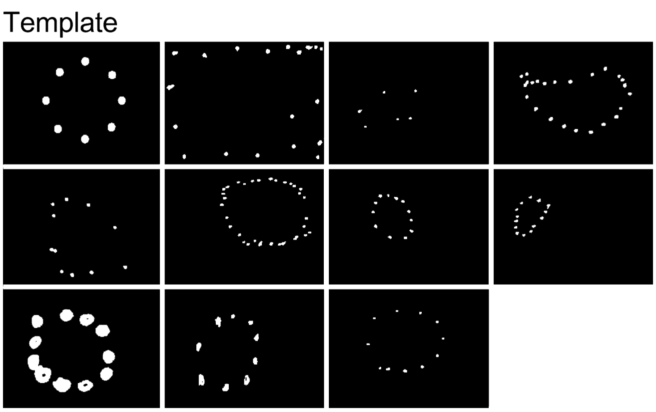

Fig. 5 Copies of a circle composed of *small, filled circles*. Arrangement is the same as in Fig. 3

score of the WS subjects was slightly but systematically higher than that of the controls, when the pairing was taken into account (mean difference=1.7, p=0.0003, paired t-test, N=10 pairs; range of scores=9–24). With respect to the verbal KBIT score, there was neither a significant correlation (p=0.46) nor a significant difference (p=0.44, paired t-test) between the two groups.

Table 1 Pairwise Spearman correlations between raters. All correlations were statistically highly significant (p<0.0001).N=3 groups × 10 subjects × 6 figures = 180 per correlation (left/right arrangements were averaged; see "Materials and methods")

	Rater 1	Rater 2	Rater 3
Rater 1	1	-	-
Rater 2	0.810	1	-
Rater 3	0.768	0.759	1

Association mKBIT scores and chronological age

There was a highly significant positive correlation between the mKBIT score and chronological age in the control subjects (r=0.917, p=0.0002, N=10 subjects) but not in WS subjects (r=0.041, p=0.91).

Analysis of copies

General

The visual templates used are shown in Fig. 1, and examples of drawings are shown in Figs. 3, 4, and 5 (control subjects) and Figs. 6, 7, and 8 (WS subjects). We illustrate copies of a simple geometrical shape (triangle; Figs. 3 and 6), copies of two connected shapes (bracket and circle; Figs. 4 and 7), and copies of a circle made of distinct, small, filled circles (Figs. 5 and 8). The drawings of all ten subjects in each group are shown, and are displayed in rows in ascending order of chronological age, from left to right. The age is given in months in Figs. 3 and 6; the copies in the remaining figures are arranged in the same order. It can be seen that subjects from groups drew comparable copies. However, three WS subjects drew only a continuous line when copying figures composed of distinct circles (Fig. 8), and one subject drew small circles connected by a line. In addition, two of these WS subjects drew a continuous line when copying the triangle made of small circles (see Fig. 1). By contrast, no control subject drew continuous lines when copying these figures. Finally, it can be appreciated qualitatively that copies improved with age in the control group but not obviously so in the WS group. This relation is quantified below.

Similarity ratings

We analyzed the similarity ratings of the copies of control subjects with respect to the template (CT), the copies of WS subjects with respect to the template (WST), and the copies of the control subjects with respect to the corresponding ones of the WS subjects (CWS) in a pair. The ratings of the three raters were highly correlated (Table 1) and did not differ significantly (p=0.78; F test in ANOVA); therefore, they were pooled and averaged per figure and subject for further analyses below. The mean ratings are depicted graphically in Fig. 9 and their values

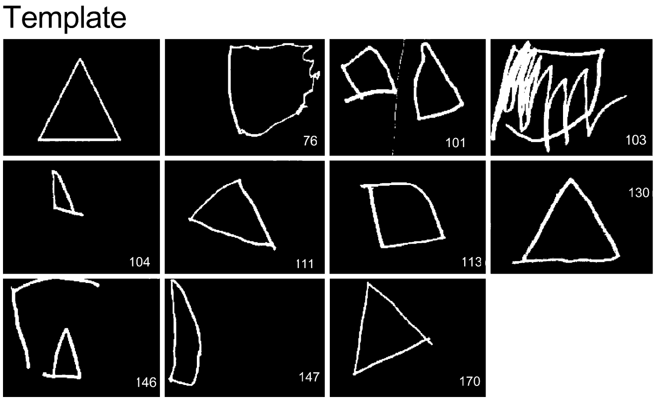

Fig. 6 Copies of the triangle by ten WS subjects. *Numbers* are biological age in months

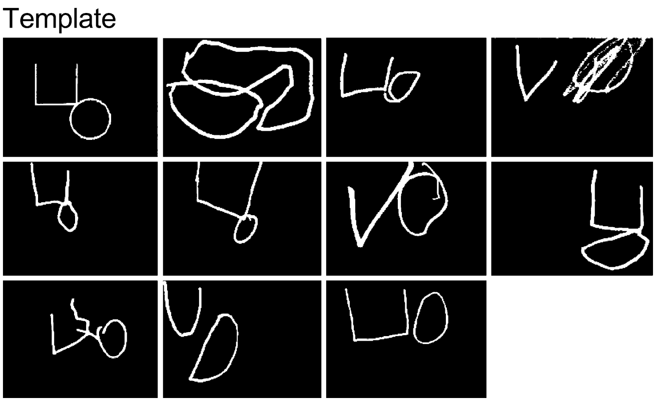

Fig. 7 Copies of another figure which is composed of two elementary shapes. Copies are arranged in the same sequence, with respect to subjects, as in Fig. 6

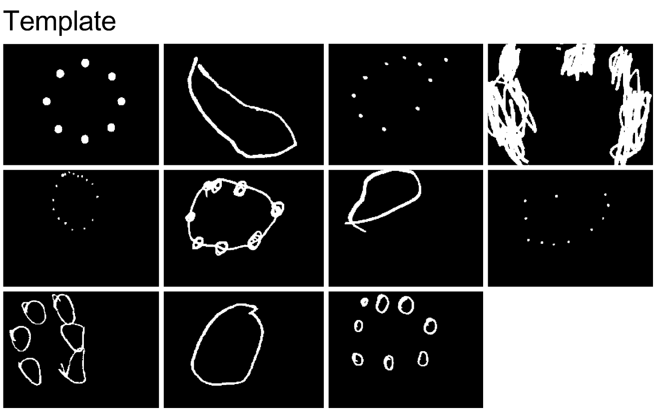

Fig. 8 Copies of the circle composed of *small, filled circles*. Arrangement is the same as in Fig. 6

and SEM are given in Table 2. The overall average similarity rating was (mean ± SEM) 54.02±3.38, 46.05 ±3.45, and 38.57±3.2 for the CT, WST, and CWS ratings (N=10 subjects × 6 figures = 60 per group), respectively. Since control and WS were paired, the effects of specific factors on the similarity rating were assessed using a repeated measures analysis of variance, as follows. The Group was a within-subjects (i.e. pairs) factor, the Figure was a between-subjects factor, and the averaged (per pair)

Table 2 Mean (SEM) ratings per figure and group (N=10 ratings per cell)

Figure	Control	WS	CWS
1	50.6 (10.6)	45.9 (9.9)	38.9 (9.6)
2	61.8 (7.3)	53.8 (7.8)	50.7 (8.4)
3	55.4 (8.8)	59.7 (9.0)	44.6 (9.4)
4	38.9 (7.8)	28.5 (6.7)	29.2 (6.5)
5	52.0 (8.2)	44.2 (8.6)	33.2 (5.9)
6	65.4 (5.6)	44.2 (6.8)	34.8 (6.3)

KBIT matrices score was a covariate. The Group factor was not statistically significant (p=0.153, Greenhouse-Geisser test); Figure was statistically significant (p=0.011, F-test) as was the matrices score ($p<10^{-10}$, F-test); finally, the Group × Figure interaction was not significant (p=0.34, Greenhouse-Geisser test).

Finally, with respect to individual templates, control subjects had overall better ratings except for one template: the intersecting lines, where WS subjects had a better score. This finding is analyzed further below.

Relation between goodness of copies and mental age

The individual mean ratings per group and figure are given in Table 2 and are displayed graphically in Fig. 9. The length of the lines are proportional to the rating and the color identifies the comparison (green for control subject vs. template, purple for WS vs. template, and red for control vs. WS rating). It can be appreciated from Fig. 9 that the green and purple lines (i.e. the ratings of the drawings vs. templates) became longer overall with mental age, but also that this tendency varied from figure to figure. The issue of the overall fit was investigated by performing a regression analysis between the ratings and mKBIT score across all figures and subjects, separately for the control, WS, and CWS ratings. The following regression model was evaluated:

$$g = k + bx + e$$

where g is the rating, k is a constant, b is the slope, x is the mKBIT score, and e is an error term. We found a significant relation between the rating and mental age. The effect of mental age was stronger and more highly statistically significant in the control than the WS group (Control: b=5.45, r^2=0.582, $p<10^{-11}$; WS: b=3.52, r^2=0.196, p=0.0004). Similarly, there was a significant relation between CWS and the average (between control and WS) mKBIT score (b=3.47, r^2=0.395, $p<10^{-7}$). This means that the drawings of matched control and WS subjects were more similar for subjects with higher mKBIT scores.

Next, we carried out a more detailed analysis with respect to individual figures. For that purpose, the regression model above was evaluated separately for

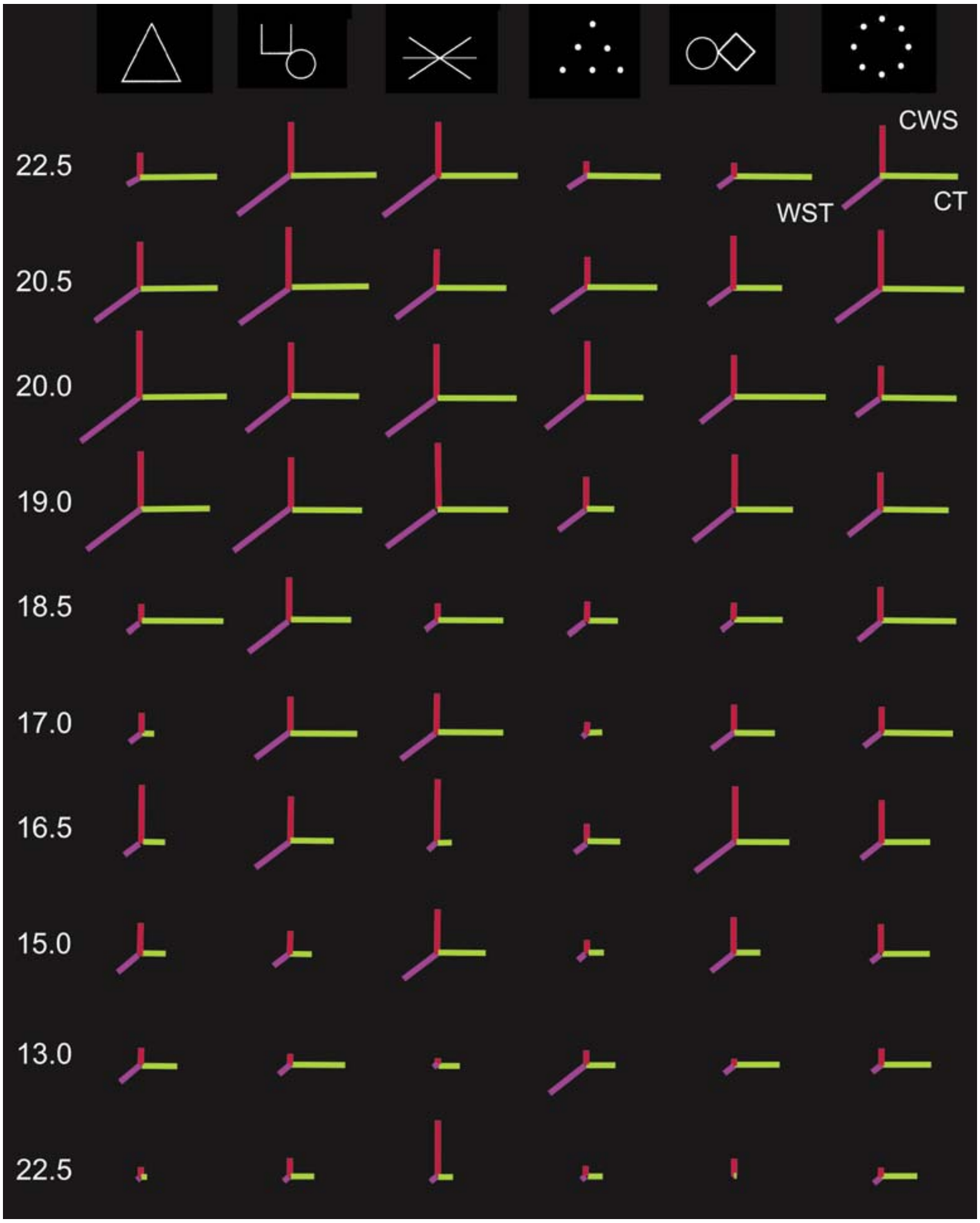

Fig. 9 Graphical representation of the goodness of copy by figure and subject. Each *triplet of lines* corresponds to a combination of a figure, a control, subject, and the matched WS subject. *Each line* is proportional to the rating; *green, purple and red lines* indicate ratings of control drawings (with respect to the template), ratings of WS drawings (with respect to the template), and ratings of similarity between corresponding control and WS drawings, respectively. *Rows* represent the ten pairs, ordered from low to high mKBIT score, bottom up. The *numbers on the left* are the average (between the control and WS pair) mKBIT score

each figure within each group of subjects. The results are shown in Table 3. We found the following. In the control group (Fig. 10), a highly significant, positive relation was found for each one of the six figures. The highest slope was observed for the simplest linear figure (triangle) whereas the two smallest slopes were observed for the two figures (triangle and circle) which were composed of small, filled circles. By contrast, in the WS group (Fig. 11), a significant relation was found only for two figures. However, these two figures were very interesting in that they were rather complex: one was a composite one (template 2) whereas the other consisted of small filled circles making up a large circle (template 6). In addition, it is noteworthy that the lack of significant correlation of the goodness of copy with mental age in the WS group was apparently due to different reasons, depending on the template. For template 4, the ratings were mostly low (floor effect); for templates 1 and 5, the ratings ranged widely but not systematically with mental age; whereas for template 3, they were mostly high (ceiling effect). These

Table 3 Slopes (and r^2, p values of t-test for slope) of the rating vs. mKBIT score relation λ for individual figures. *NS* not significant. N=10 subjects (matched pairs for CWS)

Figure	Control	WS
1	7.19 (0.672, 0.004)	3.08 (0.118, NS)
2	6.49 (0.484, 0.005)	6.09 (0.75, 0.001)
3	6.11 (0.703, 0.002)	2.18 (0.072, NS)
4	4.97 (0.604, 0.008)	2.52 (0.156, NS)
5	5.47 (0.653, 0.005)	1.97 (0.064, NS)
6	4.15 (0.821, 0.0003)	5.28 (0.737, 0.001)

findings underscore the complexity of the effects and analyzed further below.

Comparison of control and WS copies

The red lines in Fig. 9 are proportional to the similarity rating of the control vs. WS copies. It can be seen that, when the goodness of copy of a control and its paired WS subject is high with respect to the template (i.e. when both the green and purple lines are long), the corresponding copies of the control and WS subjects in a pair are also very similar (i.e. the red line is long too). This was obviously expected. It is also expected that, when the green and purple lines are of substantially unequal length, the red line should be short (e.g. cases in the top row for 4th and 5th templates): when one drawing resembles well the template and the other does not, it follows that these two drawings will be dissimilar. Now, the interesting case is when neither of the two (control and WS) copies resemble well the template. In this case, the similarity between themselves could provide useful information with respect to the WS drawing: if that similarity is high, it would indicate that both the control and WS copies differ from the template in a similar fashion; in contrast, if that similarity is low, this would indicate that the control and WS copies deviate from the template in different ways, and this could point to a potentially systematic difference of the WS copy from the corresponding control copy. We explored this issue further, as follows. First, we identified three major factors that are involved: (1) the overall rating of similarity to the template, computed as the average of the ratings of the control and WS in a pair, with respect to the template: T=(CT+WST)/2; (2) the computed dissimilarity between the control vs. template (green lines) and WS vs. template (purple lines) ratings, calculated as the absolute difference between these two ratings in a pair: CWT=|CT−WST|; and (3) the perceived dissimilarity between the control and WS copies (red lines in Fig. 9), re-expressed as DCWS=100−CWS. The data were then plotted in a 3-D plot with axes T, DCWS and CWT, shown in Fig. 12. It can be seen that three broad groups can be distinguished, roughly demarcated by the levels of T=30 and T=50. At the low end (T <30), where the overall goodness of copies is low, both the computed (CWT) and the perceived (DCWS) dissimilarity between the control and the WS copies are high. This is not surprising, given the early stage of development and the rather unstructured copies; such copies could very well differ from the template as well as between themselves. At the high end (T >60), where the overall goodness of copies is high, both CWT and DCWS are fairly low, and this is expected since copies that resemble well the template are very likely to be similar too. Now, the most interesting case is the middle group (30=T=60, in red color), where the goodness of copies is at a midrange, which, in turn, allows for differences or similarities between the control and WS copies to be more clearly manifested. Specifically, if the control and WS copies are very similar, this would indicate that they probably differ from the template in similar ways, which, in turn, would speak for a deficit in WS resulting from arrested development. By contrast, if the control and WS in this group differ appreciably from each other, this would indicate that the drawings might differ from the template in some systematic way, which would speak for "deviant" deficit. It can be seen in Fig. 12 that most of the points were at high dissimilarity levels with respect to both the CWT and the DCWS. This finding

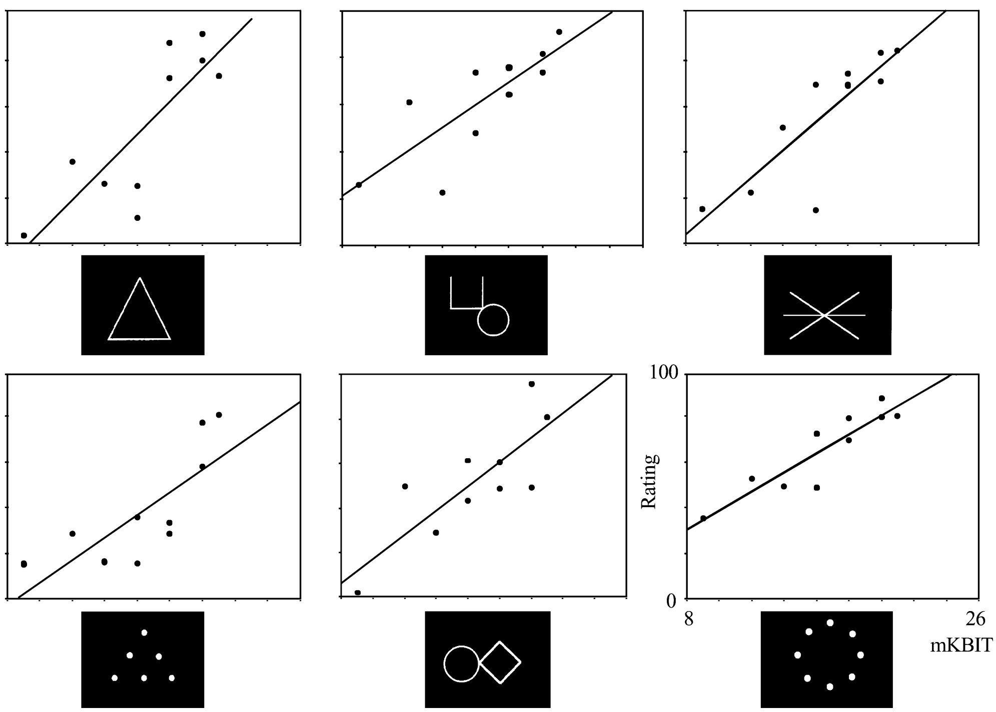

Fig. 10 The goodness of copy ratings by the control subjects are plotted against the mKBIT score for each figure. *Lines* are statistically significant linear fits (Table 3)

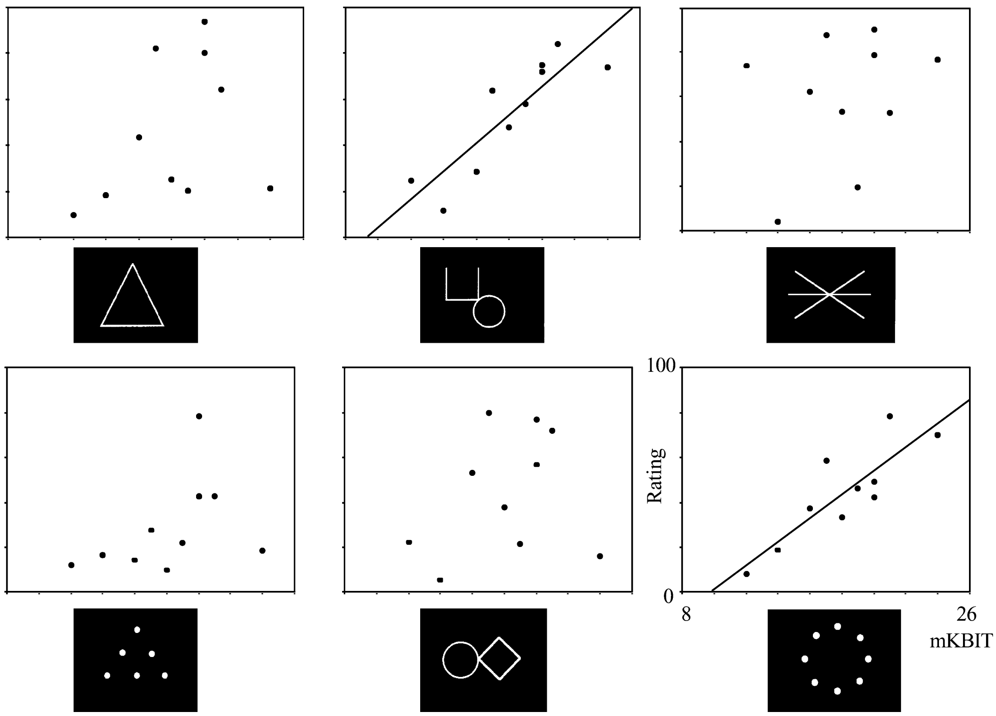

Fig. 11 The goodness of copy ratings by the WS subjects are plotted against mKBIT score for each figure. *Lines* are fitted for those cases (N=2) in which a statistically significant slope was found (Table 3)

suggests that, as development progresses and the goodness of copy improves, its relation to the template diverges between the control and WS groups, before it attains close proximity to it at a later stage in development, when the goodness of all copies is quite high. We investigated the copies during this transient period in more detail, in an attempt to identify and delineate their divergent characteristics. We identified two striking differences between the drawings of the control and the WS groups. The first was present in copies of the 3rd template depicting three intersecting lines. In this case, WS subjects typically drew three continuous intersecting lines; in contrast, the most of the control subjects drew essentially six lines emanating from a central spot (Fig. 13). Interestingly, the differences between these copies are typical of differences seen in normal development: Younger children tend to copy the model as six independent segments, whereas older children tend to copy it as three intersecting lines. This pattern also appears in tasks where perceptual matching, but not copying, is required (Stiles and Tada 1996). The second difference was present most clearly in the 6th template in which the overall shape of the circle was made of small filled circles. In this case, several WS subjects drew a solid line around the component circles whereas none of the control subject did so. Both of these findings indicate a very different perceptual organization of these templates by the two groups and in both cases it seems that the perception of the WS subjects was dominated by the overall shape, whether it was composed of intersecting lines or of small circles. By contrast, the perception of control subjects was apparently dominated by a center at the intersection of the lines. Interestingly, then, both groups added something new to the templates, namely an absent center (by the control subjects on the intersecting lines) and an absent outline (by the WS subjects on the fragmented circle; see Fig. 8).

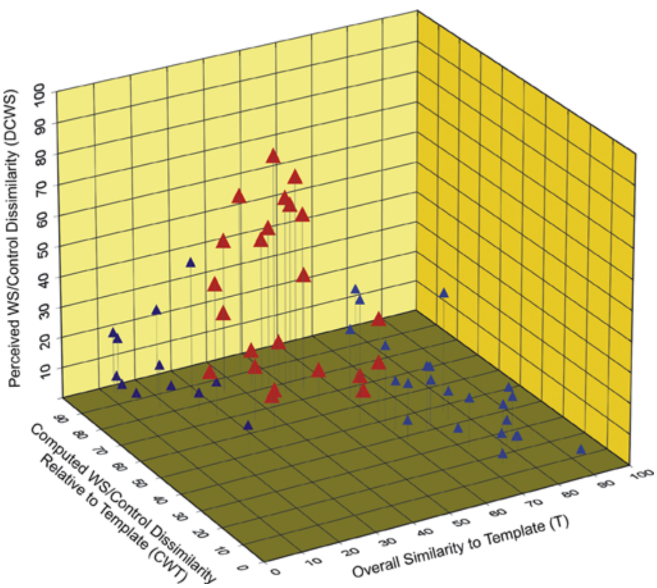

Fig. 12 Graphical display of the pairwise analysis of the matched WS and control copies. The 3-D plot illustrates the relations between the perceived dissimilarity between WS and control copies (*DCWS*) as a function of the computed dissimilarity of each drawing to its template (*CWT*) and their overall similarity to the template. *Red triangles* are ratings with an intermediate (ratings 30–60) overall similarity to the template. See text for more details

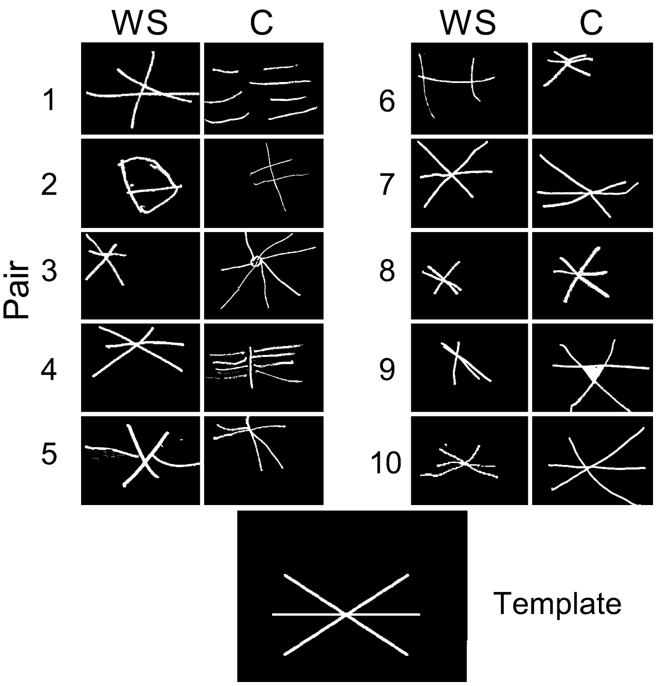

Fig. 13 Copies of the three intersecting lines (template 3) by matched WS and control subjects to illustrate the frequent fragmentation of the lines in the control drawing near their point of intersection

Discussion

The normal development of copying with age is usually subsumed under the development of constructional abilities. These abilities develop between 3 and 5 years of age, with the fastest development occurring between 4 and 5 years (Del Giudice et al. 2000). Interestingly, the developmental course of the constructional abilities is highly correlated with those of visuospatial and representational abilities, a phenomenon taken to indicate the crucial role of the latter on the former (Del Giudice et al. 2000). In general, certain figures can be copied at an earlier stage than others; for example, a circle can be copied at 3, a square at 4, a triangle at 5, and a diamond at 7 years of age (quoted in Naeli and Harris 1976). Copying, like other spatial constructional activities, involves a highly complex set of capacities that must be properly sequenced over time. These capacities include visual analysis of objects (i.e. the ability to detect features and their spatial configuration), the representation of spatial relationships among objects, and the re-creation of these in a separate space (which further requires executive processes controlling the sequencing of actions). Detailed analyses of eye-movements during block construction tasks show that WS children suffer from failure to accurately represent spatial configurations within and between blocks, rather than failure of executive processes (Hoffman et al. 2003). It is also likely that defective short- and long-term visuospatial memory (Vicari et al. 1999) as well as delayed development of fine motor skills (Plissart and Fryns 1999) could play a role in disordered copying. Because copying can be improved by appropriate training (Rand 1973), it would be interesting to evaluate the effect of such training on the copying performance in WS.

A major finding of our study was that the copies of WS subjects were very similar to those of much younger children, matched for mental age. This finding is similar to those reported by others (Bertrand et al. 1997; Mervis and Klein-Tasman 2000), and suggests that the copying deficit in WS reflects a developmental rather than a deviance disorder. This finding indicates that the copying deficit in WS reflects arrested development of this function rather than a disordered function itself. This is in accord with the observation that other cognitive functions are similarly arrested in adult WS subjects (Howlin et al. 1998), with functioning being around a 6–8 year age equivalent. It is also consistent with the finding that in our WS group, mental age (either the verbal or mKBIT score) did not improve with chronological age (range 94 months), whereas it increased significantly in the control group (range 33 months). An improvement in KBIT test performance with chronological age has been described for larger WS populations (see Mervis and Klein-Tasman 2000). The lack of such a relation in our study is likely to be due to the small sample size and, probably, to the fact that we used only the simpler vKBIT subtest (see "Materials and methods").

Our results are collectively consistent with the idea that the WS subjects showed an arrested development in all tests, including copying, at about the age of the control group. However, these conclusions have to be qualified by the finding that substantial improvement in copying by the WS subjects was observed for two complex figures (Fig. 11), which underscores the potential for improvement, in accord with the findings of other studies (Mervis et al. 1999). Moreover, the consistently superior performance of the WS subjects in copying the template of intersecting lines needs to be mentioned. This was in contrast to the copies of this template by the control subjects who tended to treat the three intersecting lines as six half-lines connected at the point of intersection (Stiles and Tada 1996). Altogether, these findings support the notion that WS is basically a developmental and not a deviance disorder (Bertrand et al. 1997; Mervis and Klein-Tasman 2000). What we mean by this is that the qualitative nature of the copying output is quite similar to that of normally developing children, albeit at younger ages. Our studies do not address the very thorny issue of whether the same result or output could emerge via different developmental mechanisms. We speculate, however, that drawing/copying—like many other visual and cognitive tasks—must draw on mechanisms that must have enormous constraints in order to generate such similarity across groups of subjects who are otherwise so different.

It is useful to compare the results of our study to those obtained in a study of subjects with Down syndrome (Clements and Barrett 1994). In both studies, subjects with disabilities were matched for mental age with healthy children, but the objects copied by each group differed appreciably. In the Clements and Barrett (1994) study, children were asked to draw partially occluded objects and to depict the occlusion. This is in contrast to our study, where subjects copied single unoccluded objects. Although the results of the two studies may not be exactly comparable, they are worth discussing. First, the drawings of subjects with Down syndrome were significantly worse than those of healthy people, whereas the drawings of people with WS in the present study did not differ significantly from those of the controls. Second, in both studies the performance of control children was significantly correlated with verbal mental age, but this was not the case for subjects with Williams or Down syndrome. Third, qualitative, systematic differences were found between the drawings of subjects with Down syndrome and those of the controls, but no obvious qualitative differences were observed in the present study, with the exception of the tendency by some WS subjects to draw continuous lines in addition to (or instead of) elements of a shape (see, e.g., Fig. 8). This latter observation is contrary to what has been previously described, namely that subjects with WS tend to copy the details of a figure while ignoring the overall shape (Bellugi et al. 2000). However, Bellugi et al.'s studies did not include detailed analyses of WS drawings compared to those of normally developing children. Bertrand et al. (1997) did compare drawings of WS children to normally developing children and found, as we did, that the drawings of WS children resembled those of normally developing children at a much earlier developmental point.

Finally, with respect to the specific locus of brain damage resulting in the abnormal copying in WS, structural MRI and postmortem studies have showed a loss of brain volume that may disproportionately involve the parieto-occipital region (Reiss et al. 2000). A detailed study of the drawing abilities of children with congenital unilateral brain lesions (Stiles-Davis et al. 1988), including copying of geometrical shapes and free drawing, has demonstrated the crucial role of the right parietal hemisphere for the normal development of these functions, as well as the lack of an obvious effect of left hemispheric lesions on these same functions. Although it is tempting to hypothesize that the right hemisphere may be involved in Williams syndrome, there is no evidence, currently, to support this hypothesis.

Acknowledgements We thank Areti Georgopoulos for conducting some of the psychophysical rating experiments, and Bagrat Amirikian and Elissaios Karageorgiou for valuable advice concerning the comparison of WS and control drawings. This work was supported by the March of Dimes grants 12-FY 97-0670, 12-FY98-0194, and 12-FY01-87 and National Science Foundation grant BCS 9808585 and 0117744 to B.L., the US Department of Veterans Affairs, and the American Legion Brain Sciences Chair.

References

Beery KE (1997) The Beery-Buktenica developmental test of visual-motor integration, 4th edn. Modern Curriculum Press, Parsippany, NJ

Behrmann M, Winocur G, Moscovitch M (1992) Dissociation between mental imagery and object recognition in a brain-damaged patient. Nature 359:636–637

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. J Cogn Neurosci 12:7–29

Bertrand J, Mervis CB, Eisenberg JD (1997) Drawing by children with Williams syndrome: a developmental perspective. Dev Neuropsychol 13:41–67

Cipolotti L, Denes G (1989) When a patient can write but not copy: report of a single case. Cortex 25:331–337

Clements W, Barrett M (1994) The drawings of children and young people with Down's syndrome: a case of delay or difference? Br J Educat Psychol 64:441–452

De Ajuriaguerra J, Stambak M (1969) Developmental dyspraxia and psychomotor disorders. In: Vinken P, Bruyn G (eds) Handbook of clinical neurology, vol 4. Elsevier, Amsterdam, pp 443–464

Del Giudice E, Grossi D, Angelini R, Crisanti AF, Latte F, Fragasi, NA, Trojano L (2000) Spatial cognition in children. I. Development of drawing-related (visuospatial and constructional) abilities in preschool and early school years. Brain Dev 22:362–367

Dryden IL, Mardia KV (1998) Statistical shape analysis. Wiley, New York, NY

Gainotti G (1985) Constructional apraxia. In: Frederiks JAM (ed) Handbook of clinical neurology. Elsevier, Amsterdam, pp 491–506

Grossi D, Orsini A, Modafferi A, Liotti M (1986) Visuoimaginal constructional apraxia: on a case of selective deficit of imagery. Brain Cogn 5:255–267

Hoffman JE, Landau B (2000) Spared object recognition with profound spatial deficits: evidence from children with Williams syndrome. Poster presented at the Cognitive Neuroscience Society, San Francisco

Hoffman JE, Landau B, Pagani B (2003) Spatial breakdown in spatial construction: evidence from eye fixations in children with Williams syndrome. Cogn Psychol 46:260–301

Howlin P, Davies M, Udwin O (1998) Cognitive functioning in adults with Williams syndrome. J Child Psychol Psychiat 39:183–189

Jordan H, Reiss J, Hoffman JE, Landau B (2002) Intact perception of biological motion in the face of profound spatial deficits: Williams syndrome. Psychol Sci 13:162–167

Kaufman A, Kaufman N (1990) Kaufman Brief Intelligence Test. American Guidance Service, Inc., United States

Kleist K (1934) Gehirnpathologie. Barth, Leipzig

Landau B, Zukowski A (2003) Objects, motions and paths: spatial language of children with Williams syndrome. Dev Neuropsychol 23:105–138

Mervis CB, Klein-Tasman BP (2000) Williams syndrome: cognition, personality, and adaptive behavior. Mental Ret Dev Disabil Res Rev 6:148–158

Mervis CB, Morris CA, Bertrand J, et al. (1999) Williams syndrome: findings from an integrated program of research. In: Tager-Flusberg H (ed) Neurodevelopmental disorders. MIT Press, Cambridge, MA, pp 65–110

Naeli H, Harris PL (1976) Orientation of the diamond and the square. Perception 5:73–77

Plissart L, Fryns JP (1999) Early development (5 to 48 months) in Williams syndrome. A study of 14 children. Genet Counsel 10:151–156

Rand CW (1973) Copying in drawing: the importance of adequate visual analysis versus the ability to utilize drawing rules. Child Dev 44:47–53

Reiss AL, Eliez S, Schmitt JE, Straus E, Lai Z, Jones W, Bellugi U (2000) Neuroanatomy of Williams syndrome: a high-resolution MRI study. J Cogn Neurosci 12:65–73

Servos P, Goodale MA (1995) Preserved visual imagery in visual form agnosia. Neuropsychologia 33:1383–1394

Snedecor GW, Cochran WG (1989) Statistical methods, 8th edn. The Iowa State University Press, Ames, IA

Stiles JU, Tada WL (1996) Developmental change in children's analysis of spatial patterns. Dev Psychol 32:951–970

Stiles-Davis J, Janowsky J, Engel M, Nass R (1988) Drawing ability in four young children with congenital unilateral brain lesions. Neuropsychologia 26:359–371

Vicari S, Brizzolara D, Carlesimo GA, Pezzini G, Volterra V (1999) Memory abilities in children with Williams syndrome. Cortex 32:503–514