Psychologica Belgica
2006, 46-4, 269-281.

LOCAL-GLOBAL VISUAL DEFICITS IN WILLIAMS SYNDROME: STIMULUS PRESENCE CONTRIBUTES TO DIMINISHED PERFORMANCE ON IMAGE-REPRODUCTION

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Impairments in visuospatial processing exhibited by individuals with Williams Syndrome (WS) have been ascribed to a local processing bias. The imprecise specification of this local bias hypothesis has led to contradictions between different accounts of the visuospatial deficits in WS. We present two experiments investigating visual processing of geometric Navon stimuli by children with WS. The first experiment examined image reproduction in a visuoconstruction task and the second experiment explored the effect of manipulating global salience on recognition of visual stimuli by varying the density of local elements possessed by the stimuli. In the visuoconstruction task, the children with WS did show a local bias with respect to controls, but only when the target being copied was present; when drawing from memory, subjects with WS produced a heterogeneous pattern of answers. In the recognition task, children with WS exhibited the same sensitivity to global figures as matched controls, confirming previous findings in which no local bias in perception was found in WS subjects. We propose that subjects with WS are unable to disengage their attention from local elements during the planning stage of image reproduction (a visual-conflict hypothesis).

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This work was supported in part by a POCTI Framework grant SFRH/BD/6020/2001 from the *Fundação para a Ciência e Tecnologia* of Portugal to Ana Maria Abreu, by Framework 6 European Commission Grant NEST 516542 to Robert M. French, and to Rosemary A. Cowell, and by CNRS funding to Scania de Schonen.

The authors would like to thank the participants and their families for their generous participation and the British Williams Syndrome Foundation for their invaluable support. Particular thanks to Francesca Happé, Christopher Jarrold and Michael Thomas for their invaluable remarks at the earlier stages of this investigation. Finally thanks to Dagmara Annaz, Maud Jacquet and Christophe Labiouse.

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Introduction

Williams Syndrome (WS) is a rare genetic disorder characterised by a distinct cognitive profile of dissociated strengths and weaknesses, and a unique social behaviour. WS results from a micro-deletion in chromosome 7q11.23 that codes the elastin gene and LIM-Kinase 1. Elastin is an essential component of connective tissue and a deletion of this gene leads to vascular disease due to the loss of some elastic properties. LIM-Kinase 1, on the other hand, is expressed in the brain and is thought to be linked to the deficits in visuospatial processing observed in WS. Genetic diagnosis for WS is obtained using Fluorescent *in situ* Hybridisation (FISH test). The FISH test is a diagnostic test of the DNA that detects the elastin deletion on one copy of chromosome 7. WS does not produce a uniform cognitive deficit, but rather clusters of deficits in visual processing (Atkinson, Anker, Braddick, Nokes, Mason, & Braddick, 2001), and relative strengths in auditory rote memory and language (for a review see, for example, Bellugi, Lichtenberger, Jones, Lai, & St George, 2000). Individuals with WS show competence in areas such as language and music. Despite the fact that they usually score lower on standardised IQ tests, they show competence in interpersonal relations. The presence of visuospatial deficits in WS has not been systematically reported for all stages of visual processing from encoding to retrieval. However, deficits in visuoconstructive tasks in subjects with WS are systematically found (e.g., Atkinson, Braddick, Anker, Curran, Andrew, Wattam-Bell, & Braddick, 2003).

One consistently reported finding in the literature concerning visuospatial cognition in WS is that such individuals possess a local bias that leads to deficits in visuospatial construction tasks. Indeed, people with WS preferentially process the component parts of global forms rather than the global forms themselves (although they are able to focus on the global level when primed to do so (Farran, Jarrold, & Gathercole, 2003)). Different hypotheses have been put forward to explain this global-local visual processing deficit found in WS.

One possible explanation is the local-bias hypothesis (e.g., Bihrle, Bellugi, Delis, & Marks, 1989), whereby individuals with WS tend to focus on local details and fail to process global visual forms adequately. Pani, Mervis, and Robinson (1999), showed, using the block design task, that performance of individuals with WS does not differ significantly from that of controls in organizing displays at the global level. The authors suggest that the problem in visuospatial construction tasks does not lie in a local processing preference, but rather in a difficulty in changing between the local and global levels of organisation. Farran, Jarrold, and Gathercole (2001) also found that children with WS process local and global information in the same way as typically developing (TD) controls; the authors suggested that their poor performance in block construction might be due to difficulty in using mental imagery.

Short Term Memory (STM) deficits have also been proposed as an explanation of deficits in visuospatial production tasks in people with WS. Although imagery and attention disorders have been suggested in an attempt to explain deficits in visuoconstructive tasks (e.g., Farran et al., 2001; Mervis, Morris, Klein-Tasman, Bertrand, Kwitny, Appelbaum, & Rice, 2003), an alternative hypothesis explains these data in terms of a visual short term memory deficit in individuals with WS (Jarrold, Baddeley, & Hewes, 1999). Specifically, Vicari, Belluci, and Carlesimo (2003) demonstrated that, in children with WS, form information is relatively preserved in STM whereas location information is not processed normally. These findings would explain Farran et al.'s (2003) demonstration of a selective impairment in construction tasks.

Fayasse and Thibaut (2002) hypothesised that, in visuoconstruction tasks, difficulty in processing the global form might stem from an inability to disengage attention from local features. If there is a tendency to over-focus attention on local elements, then increasing the number of local constituents of a global figure should impair global recognition rather than render the global form more salient. We address this issue in Experiment 2.

The general purpose of the present study is to clarify the extent to which the local bias that leads to visuospatial deficits is present in WS at different stages of visual processing and whether it depends on other cognitive components, such as memory, or on task constraints, such as Gestalt strength. The participants were children. For consistency with previous empirical work in this area, we used Navon-type stimuli (Navon, 1977). We used geometrical figures such as circles, squares, and triangles instead of letters. We chose these three simple shapes, because they are present in everyday situations (e.g., bidimensional representations of balls, boxes, rooftops, etc.), thus all the children were familiar with them.

More specifically, we address the following questions:

i. In the case of impaired production of global forms, could the memory deficits suggested by Vicary et al. (2003) explain the local bias found in some drawings by subjects with WS? In Experiment 1, we address the influence of memory in figure reproduction in WS by manipulating target presence, in order to introduce a working memory component. More specifically, two different reproduction tasks were analysed, namely, a direct copy task and a deferred memory task. Performance on these two tasks by children with WS and TD children was compared to examine the influence of memory on drawing reproduction in individuals with WS;

ii. In Experiment 2, we test the robustness of visuospatial deficits in WS under conditions of varied Gestalt strength by varying the density of local elements comprising a stimulus in a form-detection task. Fayasse and Thibaut (2002) hypothesised that WS subjects experience difficulty in disengaging attention from local features, which gives rise to deficits in processing global forms in visuoconstructive tasks. However, it is difficult to explain why an inability to disengage from local elements, which seems to be present in the performance of visuoconstruction tasks by subjects with WS, is not evident in perceptual processing in the same subjects. If this difficulty for WS subjects does indeed exist at the perceptual processing stage, then presentation of a hierarchical figure with fewer local elements should facilitate processing of the global form in subjects with WS, leading to the more frequent production of 'global' answers in a recognition task, because there would be fewer distracting local elements. Conversely, creating stimuli with fewer local elements should produce the opposite effect in TD children, because the figure is less dense and thus the Gestalt grouping law of proximity is not as strong. In Experiment 2, we compare perceptual performance of TD children and children with WS on tasks using both sparse and dense Navon-type figures, in order to address the following question: Do WS subjects experience difficulty in disengaging attention from local features in tests demanding only visual perception?

Experiment 1: Reproduction of Navon-type stimuli

In this experiment a direct copy task and a deferred memory task were administered to both children with WS and TD children, to examine the influence of short term memory on drawing reproduction in individuals with WS.

Method

Participants

This study involved a group of 13 children with WS (mean age: 8.8, *SD*: 2.24, range: 5.7-12.1 years) and a group of 22 TD children (mean age: 7.8, *SD*: 2.74, range: 4.0-12.5 years). In an attempt to match groups by mental age, younger TD children were selected.

Non-verbal ability was assessed using the Raven Coloured Progressive Matrices (CPM) (Raven, Raven, & Court, 1998). The group mean scores were '16' for the WS group (approximately corresponding to the mental age of 6 year olds) and '24' for the control group (approximately corresponding to the mental age of 8.5 years). We were not able to match children by mental-age (to obtain a one-to-one mental age match, some TD children would have to be as young as two years old and, thus, were unable to understand the tasks). Moreover, if we assume that there is a chronological maturation of the local-global processing function, then we should not compare groups with disparate chronological ages. For this reason, the TD group is younger despite presenting a higher mean mental age than the WS group. The children with WS were recruited through the Williams Syndrome Foundation, UK and all had tested positive for elastin deletion. TD children were recruited from mainstream schools. All participants were tested either at home or at school. Written informed consent was obtained by the parents of children with WS and of TD controls.

Design and procedure

Participants were required to copy a geometrical Navon-type figure (a triangle made of squares). The individual Navon-type geometric figures used as target stimuli were drawn by the experimenter for the child, in front of the child, who was then asked to reproduce the image without delay. There were two conditions: In Condition 1, the figure remained present during copying (direct-copying condition) and in Condition 2, the target figure was shown and then masked, requiring the child to copy the figure from memory (deferred-copying condition). Importantly, in the direct-copying condition, the target was available to guide the planning stages in the re-creation of the figure.

We created a rating scale to rate the children's drawing productions. The drawings were rated by using one of five different labels: "Global", "Local", "Merged", "Mixed" and "Incongruent". No response was considered correct or incorrect *per se*; our aim was to assess the degree to which responses were local or global. "Mixed", "Merged" and "Incongruent" contained both levels of responses while "Local" and "Global" contained only one level. The criteria used for rating the drawings were: respect for configural relations between the individual components to produce a recognisable global shape (Merged); the existence of an overall global form without any local components (Global); the existence of local elements making up a global form, and another global form drawn separately (i.e., a series of little squares, for example, making up a triangle and a bigger simple triangle drawn over the squares or separately – Mixed); the existence of local elements, without any global shape or intention of global shape (Local); and the existence of a merged shape with inverted forms between components and the global image, i.e., global is drawn as local and vice versa (Incongruent). Sixty-seven drawings (34 drawings from Condition 1 and 33 drawings from Condition 2 – drawings from Condition 1 and 2 by a child with WS were not rateable and another child with WS did not produce a drawing in Condition 2) produced by the clinical and control groups were rated by three independent raters, who each analysed all of the drawings. The raters were blind to the objective of the study and were not aware that the drawings had been produced in deferred and direct conditions, nor that they had been produced by TD children and children with WS. Inter-rater reliability was assessed using the Kappa statistic ($K = 0.94$).

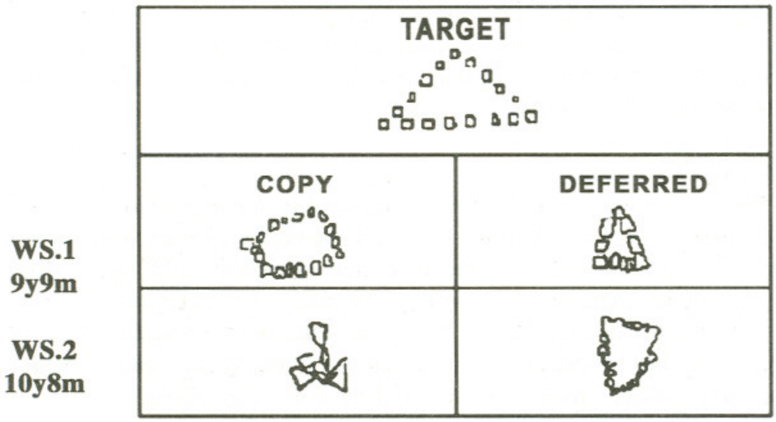

Figure 1.
Experiment 1. Representative examples of drawings by two children with WS: direct copying versus deferred-copying. Both drawings shown in the Copy condition received an 'Incongruent' rating while those deferred drawings received 'Merged' (upper drawing) and 'Mixed' (lower drawing) labels.

The results of this experiment show that the drawings of the control TD children were predominantly rated as 'Merged', indicating that they attended both to the global and to the local shape. Children in the WS group, in contrast, presented a 'local bias' in reproduction, but only when producing the drawing with the target image still in view. When drawing from memory alone, without the physical presence of the target image during copying, children with WS changed to a type of reproduction that frequently was mostly rated as 'Mixed', 'Merged', and 'Global', in striking contrast with the predominantly 'Local' productions by the same subjects in the direct-copy condition. The proportions of each type of drawing reproduction choice made by the WS group differed significantly between the 'Direct' and 'Deferred' conditions ($\chi^2(4) = 11.813$, $p = .019$). The drawing reproduction choices made by TD children, however, did not differ significantly for Direct and Deferred drawing conditions ($\chi^2(3) = .007$, $p = 1.0$).

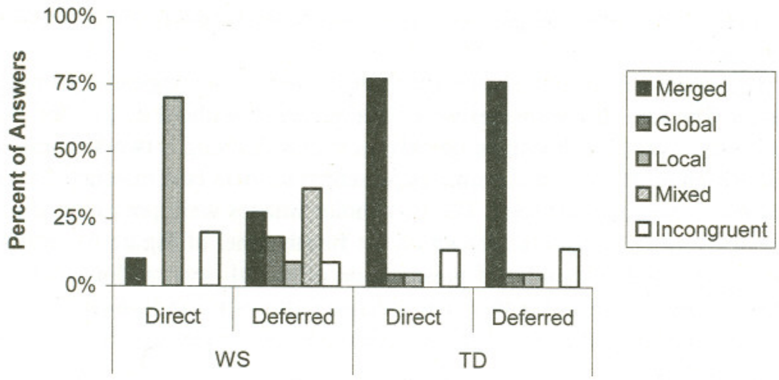

Figure 2.
Experiment 1. Drawing reproduction choices. The drawing reproduction choices do not differ between the Direct and Deferred condition for TD children. However, the drawing pattern in WS children is significantly different in the Direct condition from that produced in the Deferred condition.

Experiment 2: Perception of Navon-type stimuli (Manipulating Local-Global Salience)

If individuals with WS do possess a local bias, could more local elements composing a global shape interfere with recognition of the global form? In an attempt to answer this question, we designed a computer generated Navon-type task to assess subjects' tendencies to describe hierarchical Navon-type stimuli according to their global form by manipulating the density of local elements of the stimuli.

Method

Participants

Recruitment and details of participant groups were as described for Experiment 1.

Design and Procedure

In this experiment, participants were shown geometric Navon-type stimuli, i.e., squares, triangles and circles made up of the squares, triangles, circles, with the constraint that no figure was composed of smaller copies of itself. The task comprised 12 trials with each possible global form – square, triangle and circle – appearing twice. Each trial comprised presentation of the target stimulus, followed by disappearance of the target image and presentation of two new images: one image represented a typical "Global" answer and the other image constituted a distracter stimulus (neither Global nor Local).

The target image was shown for 900ms and then disappeared from the screen. The two choice images were then presented without delay. The children were asked to choose as quickly as possible, using a two-key answer pad, which of the two choice images presented to them best matched the target image they had just seen. The two choice images were presented simultaneously on the screen (as shown on the fourth panel of Figure 3) until the child responded. The order of presentation of stimuli was random. In 6 out of the 12 trials composing this task, a dense figure was used (made up of 24 local elements). In the other 6 trials, a sparse figure (made up of 12 local elements) was used.

The stimuli appeared on a 36 x 27 cm computer screen. They were presented on a Toshiba Satellite SM30-164 computer on a 15.4° TFT screen (1280 x 800 pixels) at a distance of ~ 40 cm from the participants. Size of hierarchical Navon-type stimuli (target image) and of simple global shapes (response stimuli) was ~ 208 x 208 pixels. Size of local shapes in the target image was ~ 32 x 32 pixels (see Figure 3).

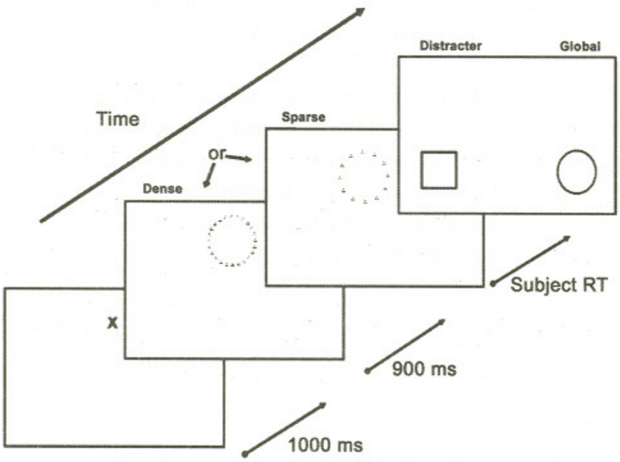

Figure 3.
Experiment 2. Computer-generated forced choice perception task to assess local global visual processing preferences with varied local component density in Navon-type geometric shapes.

Results

The results of Experiment 2 (Figure 4) indicate that manipulating the number of local elements did not have a significant main effect on the proportion of global answers ($F(1, 62) = .854$, $p = .359$). Somewhat surprisingly, there was also no significant main effect of group: Children with WS did not perform significantly differently in terms of their proportion of global answers compared to TD children ($F(1, 62) = 0.924$, $p > 0.34$). Contrary to our expectations, subjects with WS produced fewer global type answers when presented with sparse figures, and the number of global answers given by control subjects was not affected by the density of the figures presented. However, the interaction between Group-Type and Density-Type was not statistically significant: $F(1, 62) = 2.77$, $p = 0.10$.

We also considered RTs for this task. Given the learning disability usually found in subjects with WS, as evidenced by their lower scores on the CPM, it was not surprising to find significantly longer reaction times in the WS group compared to the TD group, $F(1, 62) = 7.77$, $p = .007$. The main effect of Density-Type, as well as the Group-Type X Density-Type interaction were not significant.

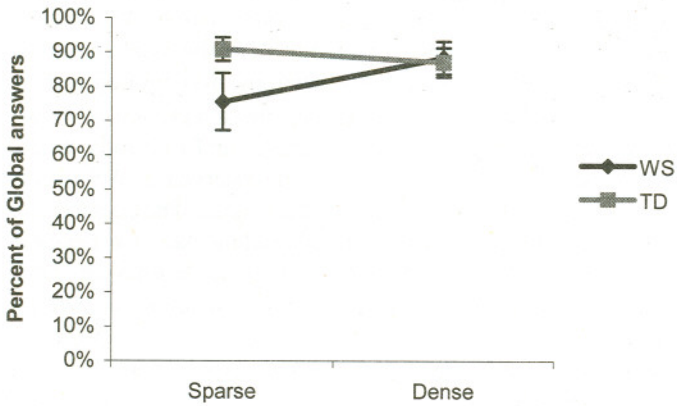

Figure 4.
Experiment 2. Graphical representation of the percent of global choices in sparse-dense manipulations made by TD and WS participants. Standard error bars are included.

Discussion

We have shown that children with WS do not behave in the same way as controls in a reproduction task. We have also shown that varying the density of the local elements of a global shape does not impact on the number of global answers given by the participants from either group: In the perception task, the participants with WS performed similarly to TD subjects. These findings corroborate Farran et al.'s (2003) suggestion that processing preferences in WS are task dependent.

The most striking finding of the present study emerged from the examination of drawing reproduction by children with WS in Experiment 1: WS children showed a strong local bias in the direct copy condition, but this local bias in WS children disappeared in the deferred copy condition. Anecdotally, we noticed a type of recurrent processing of the target by children with WS: In the direct copy task, participants with WS repeatedly referred to the target image every time they finished producing a local element. We propose a "visual conflict" hypothesis (Abreu, French, Annaz, Thomas, & de Schonen, 2005) to explain these results. We believe that three factors may be involved in the poorer performance of WS subjects in the copying task when the target remains present. These are: i) The frequent referral to the physically present image combined with an increased focus on its local elements produces a refreshing of these local components in memory, thus causing a conflict with the global image held in memory. It seems likely that this global image exists in memory, since children with WS perform well on recognition tasks; ii) Their difficulties in retaining location information (Vicari et al., 2003) mean that, even if they accurately perceive all of the local components, they cannot reproduce them at their correct locations; iii) Children with WS might have deficits in local feature binding, i.e., the integration of local features into a whole. This is congruent with findings in an EEG study, where deficits in gamma-band bursts (around 40 Hz) were observed in WS children while looking at coloured pictures of adult female faces. The gamma oscillations are thought to be associated with visual binding (see, for example, Grice, Spratling, Karmiloff-Smith, Halit, Csibra, de Haan, & Johnson, 2001; Singer, 1995). The continuous reintroduction of local visual features from the physically present image might impede the visual system from effectively settling on a binding between the local and global features in memory, resulting in the production of drawings that either neglect the global form altogether, or confuse the global form with the local form. In contrast, we suggest that the image of a figure held in working memory might more closely resemble a "Gestalt" perception with a reduced level of local detail compared to the original, physical image. This would tend to make it easier for children with WS to reproduce the image because the visual interference created by the physical presence of local components of the original image is reduced.

If subjects with WS have difficulty disengaging from local elements at the perceptual as well as the production level, decreasing the number of local elements in a perception task should promote global recognition answers by children with WS. The results of Experiment 2 demonstrated that the presentation of sparse target images did not increase, but rather tended to diminish, the number of global answers given in a recognition task by subjects with WS, although this was not statistically significant. The presentation of sparse figures had no effect on global answers given by control participants. This suggests that children with WS process hierarchical shapes in a similar manner to control children, but that they show a tendency to be more sensitive to a change in the number of local elements or to a change in the density of local elements in the global shape.

Our findings shed new light on the disparate interpretations on visual processing in WS. First, we confirm that there is no perceptual deficit even when manipulating density of local elements. Second, we substantiate previous reports on reproduction deficits in WS (e.g., Farran et al., 2001; 2003).

However, these deficits are observable only when the target remains visible. It would be interesting to further investigate visuospatial perception in conditions where the target remains visible. If manipulation of memory resources can influence local-global type responses in reproduction tasks, this implies a need for a controlled experiment where the target remains on-screen in a perception task. Such an experiment could unify the different hypotheses put forward on visual processing in WS by either providing evidence for consistent preservation of visuospatial perception or showing that the visual-conflict hypothesis is also valid for perception tasks where the target remains visible (recall that in our perception task the target was absent during response). It is probable, however, that the visual-conflict hypothesis emerges only in reproduction tasks. Thus, a new series of direct/deferred experiments, using not only drawings, but also simple block design tasks, for example, might allow the demonstration of the generalisation of the deficit to all construction tasks in direct situations. Such findings could shed new light on the previous deficits found in reproduction tasks described in the literature that only make use of direct (visible target) conditions.

Conclusion

The results of Experiment 2 indicate that children with WS perform as well as control children at recognition of hierarchical Navon-type figures. The manipulation of the density of local elements in the images presented for recognition produced no alteration in the performance of either group, suggesting the use of similar visual processing strategies. It was only in the drawing production tasks of Experiment 1 that the local bias of children with WS became evident, specifically, when the target remained present. We have suggested a "visual conflict" hypothesis to explain these results.

The "visual conflict" hypothesis is in accordance with, and integrates, several hypotheses regarding the visuoconstructive deficits seen in children with WS. These include difficulty in changing between local and global levels of organisation (Pani et al., 1999), difficulty in using mental imagery (Farran et al., 2001), inability to disengage attention from local features (Fayasse & Thibaut, 2002), and difficulty in adhering to spatial relations when integrating parts of an image (Farran et al., 2003). All of these deficits seem to be triggered only when reproduction is required. We have suggested that the deficits described in the foregoing hypotheses might be enhanced by a conflict between imagery and perception, by invoking Mervis et al.'s (2003) account of highly focused attention in children with WS. Specifically, we have suggested that each time a child with WS refers back to the target image, the child over-focuses on one single local element or on a limited number of local elements; when the child with WS returns to the drawing in progression, the global configuration of the local element or elements that the child focused on is lost in the attempt to reproduce the element adequately. We suggest that the recurrent processing of target images by children with WS should be further analysed in a controlled study to determine the contribution of this behaviour to reproduction difficulties in the presence of a target.

Our findings might contribute to the creation of adapted teaching methods that consider visual processing constraints in WS. In addition, these findings might shed new light on the profile of cognitive function in people with WS.

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Received May, 5, 2006
Revision Received October, 26, 2006
Accepted November, 13, 2006