Perceptual Grouping Ability in Williams Syndrome: Evidence for Deviant Patterns of Performance

Emily K. Farran

Department of Psychology

University of Reading

Address correspondence to:

Emily Farran
Department of Psychology
University of Reading
Earley Gate
Reading
RG6 6AL
UK
Tel: +44 (0)118 378 7531
Fax: +44 (0)118 931 6715
E-mail: E.K.Farran@reading.ac.uk

This research was funded by the Reading Endowment Trust Fund

Abstract

Williams syndrome (WS) is a rare genetic disorder. At a cognitive level, this population display poor visuo-spatial cognition when compared to verbal ability. Within the visuo-spatial domain, it is now accepted that individuals with WS are able to perceive both local and global aspects of an image, albeit at a low level. The present study examines the manner in which local elements are grouped into a global whole in WS. Fifteen individuals with WS and 15 typically developing controls, matched for non-verbal ability, were presented with a matrix of local elements and asked whether these elements were perceptually grouped horizontally or vertically. The WS group were at the same level as the control group when grouping by luminance, closure, and alignment. However, their ability to group by shape, orientation and proximity was significantly poorer than controls. This unusual profile of grouping abilities in WS suggests that these individuals do not form a global percept in a typical manner.

Perceptual Grouping Ability in Williams Syndrome: Evidence for Deviant Patterns of Performance

Introduction

Williams syndrome (WS) is a rare genetic disorder, which, amongst other characteristics, displays an unusual cognitive profile. Individuals with WS have an approximate IQ of 60, which comprises significantly higher levels of verbal compared to visuo-spatial ability (e.g. Udwin & Yule, 1991).

Visuo-spatial processing in WS has predominantly been explored in relation to the local processing bias hypothesis, i.e. a preference to process the parts of an image at the expense of attending to the global form (e.g. Bellugi, Sabo, & Vaid, 1988). Further investigation has since demonstrated that this is true for drawing and construction tasks (e.g. Bihrle, Bellugi, Delis, & Marks, 1989; Rossen, Klima, Bellugi, Bihrle & Jones, 1996), but not for perceptual tasks; the pattern of local and global processing on perceptual tasks resembles that of typically developing (TD) controls (e.g. Farran, Jarrold, & Gathercole, 2001; 2003). Given that global processing *is* available to individuals with WS, the present study aimed to determine *how* this is achieved, by examining perceptual grouping.

Perceptual grouping is the process in which local elements within a visual field are perceptually grouped together into global wholes (e.g. Kohler, 1929; Wertheimer, 1923). This was once thought to be a single mechanism. However, behavioural (e.g. Ben-Av & Sagi, 1995) and neuroanatomical (e.g. Altmann et al., 2003; Kourtzi et al., 2003) evidence from the typical population have since demonstrated differential processing across grouping types. This differentiation in processing, and evidence from the WS literature reviewed below, raise the possibility that the profile of perceptual grouping abilities in WS may be atypical.

Pani et al. (1999), in a visual search task, showed that WS and control groups were more influenced by the grouping of stimuli (grouped by ‘good form’: elements that form a regular/ predictable spatial arrangement), than the number of elements. This suggests that, as in typical development, perceptual grouping has an influence on performance in WS. Overall level of performance was significantly poorer in the WS group than controls. However, the control group were typical adults, thus it is not possible to know whether the level of performance of the WS group was commensurate with their non-verbal mental age.

Wang, Doherty, Rourke, and Bellugi (1995) employed the Gestalt Closure subtest of the Kaufman Assessment Battery for Children (Kaufman & Kaufman, 1983), the Mooney faces test (Mooney, 1957), and the anomalous contours test (Hamsher, 1978). Performance on these measures of grouping by closure (elements which form closed units) was comparable between a WS group and a Down syndrome (DS) group (Wang et al., 1995). Individuals with DS are not an ideal comparison group as they have an unusual cognitive profile (Klein & Mervis, 1999). However, performance on the Gestalt closure task was compared to norms; both groups performed within the range expected for young school-aged children, which is considerably below their chronological age (mean 15.7 years).

Grice et al. (2003) presented participants with Kanizsa squares (figures with illusory contours; Kanizsa, 1978) and measured grouping by closure behaviourally and at a neurophysiological level using ERPs. Individuals with WS could group by closure to perceive illusory contours. However, this ability was associated with deviant neural processing in the temporal-occipital areas; controls, but not individuals with WS, showed a larger N1 amplitude for illusory compared to non-illusory stimuli.

Atkinson et al. (1997) employed motion and form coherence tasks as respective measures of dorsal stream (occipital lobe to the parietal lobe) and ventral stream (occipital lobe to the inferior temporal lobe) functioning. In the motion coherence task, a proportion of elements within a target rectangle oscillate in the opposite direction to the background elements. Success relies on grouping by common fate. In the form coherence task, a proportion of line segments are arranged into concentric circles, whilst the remaining elements are randomly oriented. Success is dependent on grouping by similarity. The pattern of WS performance revealed a relative deficit in motion coherence, compared to form coherence. Overall level of ability was also poor in WS, with higher threshold values than controls on both tasks. However, controls were not matched to the WS group.

Neuroanatomical studies appear to indicate abnormalities in WS in areas implicated in perceptual grouping. The neural substrates common to all forms of perceptual grouping are early visual areas V1 and V2 (Kapadia, Westheimer, & Gilbert, 1998; Ross, Grossberg, & Mingolla, 2000). Galaburda and Bellugi (2000) reported a well-differentiated area V1 in their autopsy study of 4 WS brains. Further investigation of the layers of V1 (Galaburda, Holinger, Bellugi, & Sherman, 2002) showed abnormalities such as areas of increased cell packing and neuronal size differences in WS brains, compared to control brains.

MRI studies showed increased gyrification (cortical folding) in WS in the right parietal and occipital lobes (Schmitt et al., 2002), disproportionate reduction in parietal-occipital regions and a left dominance of occipital lobe in WS relative to controls (Reiss et al., 2000). These abnormalities are also consistent with activation during perceptual grouping in the typical population: ERP recordings have shown that grouping by proximity in the typical population activated from striate (V1) or prestriate cortex to medial occipital and parietal cortex, whilst grouping by shape similarity activated occipitotemporal areas (Han et al., 1999). Thus, although precise predictions cannot be made, brain abnormalities clearly predict that perceptual grouping in WS may be atypical.

One cannot investigate visuo-spatial perception in WS without alluding to a second predominant hypothesis within the WS literature, that this population have a dorsal stream deficit (Atkinson et al., 1997). The dorsal stream was initially thought (Ungerleider & Mishkin, 1982) to be responsible for processing spatial properties, whilst the ventral visual stream processed visual object properties. Despite the predictive value of this differentiation for perceptual grouping performance in WS, a dorsal deficit is not explored in the present study for the following reasons. As observed above, neuroanatomical support for a dorsal deficit in WS is mixed. Furthermore, Atkinson and colleagues have recognised that the circuits activated when processing motion and form coherence are not "… strictly 'dorsal' and 'ventral'…" (Braddick, Atkinson and Wattam-Bell, 2003, p. 1774, also see Braddick et al., 2001), and that weaker motion than form coherence is not specific to WS (Braddick et al., 2003). A dorsal stream deficit in WS can therefore be discounted, at least as an explanation for the characteristic WS visuo-spatial cognitive profile. Moreover, the Ungerleider and Mishkin (1982) model is no longer tenable (Milner and Goodale, 1995; Goodale and Milner, 2004). A comparison between spatial and visual grouping abilities cannot therefore inform dorsal and ventral functioning. Nevertheless, it can speak to the monolithic view of perception (Ungerleider & Mishkin, 1982).

The standard procedure for investigating perceptual grouping is to present a matrix of elements grouped into rows or columns. Investigations of the onset of perceptual grouping in infancy suggest that grouping by luminance is the most robust form of similarity grouping (Bremner, 1994); it has been shown at 3 months (Quinn, Burke, & Rush, 1993) and in newborns (Farroni, Valenza, Simion, & Umilta, 2000). In contrast, grouping by shape similarity is available at 7 months (Quinn, Bhatt, Brush, Grimes, & Sharpnack, 2002). Studies with typical adults assume that grouping which occurs in a short space of time represents a computationally simpler perceptual mechanism (Han et al, 1999; Kurylo, 1997). Grouping by proximity (elements that are close together) occurs before grouping by closure, orientation (elements of the same orientation) (Chen, 1986), luminance (Ben-Av and Sagi, 1995), shape (X and L shapes; Ben-Av and Sagi, 1995; Han et al., 1999; 2001), and alignment (Kurylo, 1997). Closure is available earlier than grouping by orientation (Chen, 1986), and there is no difference in availability between grouping by luminance and shape (Ben-Av & Sagi, 1995). One could predict that individuals with WS will show a profile which favours more robust or less computationally demanding grouping types with strengths, therefore, in grouping by luminance and/or proximity.

Behavioural and neuroanatomical studies indicate that types of grouping are operated by separate mechanisms: differentiation is observed between similarity grouping and spatial grouping, as well as within these categories. As such in the present study, four types of similarity grouping and two forms of spatial grouping were investigated in WS. Participants were shown a matrix of elements and asked whether they are grouped into rows or columns. The strength of each grouping category was manipulated systematically as each condition progressed to determine threshold levels of ability.

Method

Participants

Fifteen individuals with WS were recruited from the records of the Williams Syndrome Foundation, UK. All individuals had been positively genetically diagnosed with WS by a Fluorescent in-situ Hybridisation (FISH) test. This checks for the deletion of elastin on the long arm of chromosome 7, a deletion common to approximately 95% of individuals with WS (Lenhoff, Wang, Greenberg & Bellugi, 1997). Elastin is responsible the heart problems associated with WS. Therefore, in addition to genetic information, diagnosis is also based on phenotypic information. All 15 individuals had been clinically diagnosed using phenotypic and genetic information. The WS group were matched individually to 15 typically developing (TD) children. As the profile of abilities is not uniform in WS, it would be inappropriate to match by general mental age. Equally, if matched by verbal ability, the WS group would display inferior non-verbal performance relative to the control group. The groups were therefore matched by their score on the Ravens Coloured Progressive Matrices (RCPM; Raven, 1993). This is a recognised non-verbal measure of fluid intelligence (Woliver & Sacks, 1986) and thus gives a general measure of non-verbal ability, which also avoids the problem of matching away any group differences. Table 1 illustrates the RCPM scores, and Chronological ages of each group.

Design and Procedure

Each task was presented on a computer monitor. The individual was presented with a grid containing an arrangement of 49 elements in a 7 by 7 formation (with the exception of the proximity task, in which the number of elements varied with changes in proximity). They were asked to press one of two response buttons, which were labelled with a horizontal or vertical two-headed arrow, to indicate whether the stimuli were grouped horizontally or vertically. Stimuli remained on the screen until a correct response had been made to provide participants with feedback. The stimulus was then replaced by a 500 ms. mask before the next trial began. For both participant groups, in order to ensure that the individual could understand the procedure, the experimenter demonstrated vertical and horizontal using visual cues such as their hand or by drawing an imaginary line down the monitor screen. The participant was asked: “Do you think they (pointing at the image elements) are lined up this way (vertical visual cue) or this way (horizontal visual cue)? They were then shown which button to press to indicate their response. All children were able to grasp this procedure during the practice trials.

Grouping by similarity.

Grouping was based on four dimensions of similarity: shape, luminance, orientation, or closure. The spatial position of each element remained constant, thus grouping was defined by the visual identity of each of the elements. The four grouping categories: shape, luminance, orientation, and closure, were tested within one task, as four counterbalanced blocks of grouping type (see Figure 1).

Closure: This is not strictly a measure of grouping by closure. It is a form of grouping by shape similarity (see below), but introduces topological properties to the stimulus array, as in typical closure tasks (e.g. Kanizsa illusion; Kanizsa, 1978). The shapes used were a triangle with 3 sides depicted (closed) versus a triangle with 2 sides depicted (open).

Shape: Elements were either squares or circles, thus grouping was based on shape similarity.

Luminance: Elements were all circles. These were either black or white.

Orientation: Single lines were presented. These were either at 0 degree orientations (vertically upright) or slanted 30 degrees clockwise.

The task commenced with 8 practice trials, two from each visual category. The experimental trials were in 4 blocks, one for each grouping category. In each block, trial difficulty was increased in order to obtain threshold values. This occurred sequentially by introducing distracter elements which conflicted with the grouping of the remaining elements (see Figure 1). In each grouping block, the initial 8 trials had no distracting stimuli (level 1), the remaining 12 trials consisted of levels of difficulty in which 0 to 6 distracting elements were present, one of each grouped by rows or by columns respectively. These were divided into level 2 (1 to 3 distracting elements) and level 3 (4 to 6 distracting elements). Excluding the practice trials, there were 20 experimental trials for each grouping type. Counterbalancing was carried out by employing two different orders of block presentation, which were each presented to half of the participants. These were: closure, orientation, shape, luminance; or shape, luminance, closure, orientation. 50% of trials were grouped vertically and 50% showed horizontal grouping of elements.

Spatial grouping.

Alignment: Individuals were presented with a grid of 7 by 7 (49) unfilled circles. They were asked to indicate whether the elements were aligned in a straight line horizontally or vertically. Each circle had a diameter of 20 pixels, and when aligned, circles were spaced by a 20 pixel gap. Task difficulty increased sequentially according to the extent to which the elements were misaligned. It was ensured that along each column or row, 4 or 3 elements were misaligned in the same direction, whilst the remaining elements did not change position. Approximately 50% of the 49 elements were misaligned (24 or 25 elements). In the first 8 trials, misalignment was by 9 pixels. The data from this set is recorded as level 1. As these were the first trials, the initial 2 were disregarded as practice trials, leaving 6 trials. The remaining trials were presented in a further two blocks. Block two was the easier block, in which misalignment varied from 9 to 5 pixels, and in block three, misalignment was from 4 to1 pixels. Each increment of misalignment was displayed twice, once as a row, and once as a column, thus there were 10 trials in block two and 8 trials in block three. There were 24 experimental trials in total. These were divided into 5 levels in order to obtain threshold values; level 1, misalignment by 9 pixels (6 trials), level 2, misalignment by 9 to 7 pixels (6 trials), level 3 was by 6 to 5 pixels (4 trials), level 4, misalignment from 4 to 3 pixels (4 trials), level 5, misalignment by 2 to1 pixels (4 trials). Examples of stimuli are shown in Figure 2.

Proximity: Participants were shown a grid of unfilled circles, 20 pixels in diameter (see Figure 3). These were grouped together horizontally or vertically by proximity. Arrangements were a standard 7 circles, 20 pixels apart in one dimension, horizontal or vertical, but were more proximal in the opposing dimension. In the first 8 trials, circles were 5 pixels apart in the more proximal dimension. There were 10 circles in this dimension to maintain the overall ‘squareness’ of the arrangement. These data were scored as level 1, with the first two trials disregarded as practice trials. The remaining trials were presented in a further two blocks, each increment of proximity displayed as a column and as a row. In block two, circles were proximal by 5 to 11 pixels in one dimension. The number of circles in the more proximal dimension varied to maintain overall squareness. This was 10 circles for proximity of 5 to 7 pixels (level 2, 6 trials) and 9 circles for proximity of 8 to 11 pixels apart (level 3, 8 trials). Block 2 was harder, with proximity in one dimension by 13 to 19 pixels, compared to 20 pixels in the opposing dimension. The number of circles in the more proximal dimension was 8 circles for proximity of 13 to 15 pixels (level 4, 6 trials), and 7 circles for proximity of 16 to 19 pixels (level 5, 8 trials). There were 34 experimental trials in total.

Results

Results are reported according to thresholds values for grouping. A threshold is the point at which the grouping of stimuli, based on visual or spatial attributes, becomes apparent. Differences between stimuli below the grouping threshold are too similar across dimensions (horizontal and vertical) to elicit a grouping effect, and thus performance is at chance. In this experiment, thresholds were ascertained by observing the proportion of correct responses of each individual, at each level of difficulty. The threshold value is the difficulty level reached previous to the level at which performance is at chance (equal or less than 50% accuracy).

The performance at each threshold level, for each grouping type, was also compared to ceiling performance (100% accuracy). One-sample t-tests revealed that performance was consistently significantly different from ceiling in both groups (p<0.05 for all).

Grouping by similarity

Individuals' threshold values were analysed using a mixed design ANOVA with 2 factors; grouping type (4 levels: luminance, orientation, shape, closure), and group (2 levels: WS, TD). Results are illustrated in Figure 4. These showed a main effect of group, $F(1, 28)=10.36$, $p=0.003$, partial $\eta^2= .27$, due to higher thresholds in the WS than the TD group. There was a main effect of grouping type, $F(3, 84)=2.85$, $p=0.04$, partial $\eta^2=.09$. Paired samples t-tests revealed that this was due to superior performance in the ability to group by luminance compared to grouping by shape

($t(29)=2.06$, $p=0.05$) and closure ($t(29)=3.17$, $p=0.004$). The interaction between grouping type and group was also significant, $F(3, 84)=2.71$, $p=0.05$, partial $\eta^2=.09$. Independent samples t-tests showed that this was due to significantly poorer performance in the WS group compared to the controls in grouping by shape ($t(28)=3.00$, $p=0.01$) and orientation ($t(23.54)=3.86$, $p=0.001$). There was no group difference in the ability to group by luminance ($t(28)=1.37$, $p=0.18$) and closure ($t(28)=1.32$, $p=0.20$).

Spatial grouping

The data from one individual with WS were lost due to computer error. Therefore their matched control was taken out of the analyses, leaving 14 participants in each group. A mixed design ANOVA was carried out, with grouping task (2 levels: proximity, alignment) and group (2 levels) as factors (see Figure 5). This revealed a main effect of group, $F(1, 26)=4.99$, $p=0.03$, partial $\eta^2=.16$ (WS<TD). The main effect of grouping type was not significant, $F(1, 26)=3.11$, $p=0.09$, partial $\eta^2=.11$. There was a significant interaction between grouping type and group, $F(1, 26)=11.16$, $p=0.03$, partial $\eta^2=.17$. Independent samples t-tests revealed that this was due to a significant group difference on the proximity task only (proximity, $t(26)=2.76$, $p=0.01$, WS<TD; alignment, $t(26)=.29$, p=0.77).

Comparison across tasks

Having established that the performance of the WS group overall differed from that observed in typical development, we were interested in how this difference varied across tasks. A direct comparison of threshold values is not appropriate as it would be difficult to compare the extent to which any differences in the absolute level of ability reflected impaired or unimpaired performance. Therefore, threshold values of the WS group were transformed for each task into z-scores on the basis of the performance of controls, as this partials out any difference in the relative difficulty across the tasks that occur in typical development (see Figure 6). A one-way ANOVA was carried out on the z-scores of the WS group with one within-participant factor of grouping type (6 levels). The main effect of grouping type was significant, $F(5, 65)=5.92$, $p<.001$, partial $\eta^2=.31$. Paired t-tests revealed that this was due to significantly higher levels of ability in grouping by alignment task relative to grouping by proximity ($t(13)=2.88$, $p=0.01$), shape ($t(13)=3.99$, $p=0.002$), and orientation ($t(13)=3.85$, $p=0.002$). Grouping by closure was significantly stronger than grouping by shape ($t(13)=3.44$, $p=0.004$) and by orientation ($t(13)=3.47$, $p=0.004$). Grouping by luminance and proximity was also significantly higher than grouping by orientation (luminance and orientation, $t(13)=2.58$, $p=0.02$, proximity and orientation, $t(13)=2.32$, $p=0.04$).

Discussion

Previous studies suggest that individuals with WS can process perceptual stimuli at both local and global levels (Farran et al., 2001; 2003). The aim of this study was to investigate how global processing is achieved in WS. The WS group were matched individually to the TD controls by performance on the RCPM. One can assume, therefore, that the two groups had comparable levels of visuo-spatial ability. This enables one to determine the level of ability of the WS group on each task relative to their general level of visuo-spatial ability.

Performance on four similarity grouping tasks indicated that the WS group’s ability to group by luminance and closure was at the same level as the control group i.e. commensurate with their general level of visuo-spatial cognition. In contrast, the ability to group by shape and orientation was significantly poorer than that of the controls. Spatial grouping performance was also atypical in the WS group: whilst they were able to group by alignment at the same level as the controls, the ability to group by proximity was significantly poorer in the WS group than the control group.

The profile of performance across all tasks showed that, relative to the controls, grouping by alignment, luminance and closure were the strongest in WS, followed by grouping by proximity, then shape, and finally orientation. This demonstrates that individuals with WS do not form a global percept in a typical manner. It is therefore possible that this population rely on their stronger grouping abilities, alignment, luminance, and closure, when creating a global percept for object recognition. The use of a restricted set of grouping principles could explain why visuo-spatial perceptual abilities are poor in WS.

As discussed in the introduction, the results of this study cannot speak directly to Atkinson et al.'s (1997) suggestion of a dorsal stream deficit in WS, since the Ungerleider and Mishkin 'what' versus 'where' conception is not nowadays regarded as tenable (Milner and Goodale, 1995; Goodale and Milner, 2004). However, the present results do add support to the argument against the Underleider and Mishkin (1982) model of perception. Differentiation in grouping ability was seen in WS within both spatial and similarity domains. This is inconsistent with a generalised monolithic model, which would predict a universal deficit across a whole domain.

The ability of the WS participants to group by alignment is consistent with the results of Pani et al.'s (1999) visual search task in which individuals were able to group by a similar spatial mechanism, good form. Relative to the performance of the control group, the ability to group by alignment in WS was significantly better than grouping by proximity. This indicates deviant processing, as adult participants show patterns of spatial grouping abilities in the opposite direction, i.e. a superior ability to group by proximity compared to alignment (Kurylo, 1997). This dissociation in WS does not, therefore, support the notion that less computationally demanding types of grouping might be relatively superior in WS. However, it is possible that, in light of their poor perceptual grouping abilities overall, the WS group sought alternative strategies where possible. The alignment task has this potential as participants could observe a single row or column of the display to judge alignment, rather than the overall gestalt form. Such a strategy would not be possible on the proximity task, which could explain the relatively elevated performance of the WS group on the alignment task. It is unlikely that the control group would look for alternative strategies, as they show little difficulty in using perceptual grouping.

Differences in completion strategy do not appear to account for the differences in similarity grouping ability in WS. The relative strength in grouping by luminance is consistent with the notion that this may be one of the more robust forms of grouping (Bremner, 1994). Individuals with WS are born with atypical brain structure, thus the developmental process of their brain is also atypical. It is possible that this robust form of grouping is less vulnerable to faulty development than other forms of grouping, which would explain the relatively superior level of performance on this task in WS. In addition, if luminance grouping is present at birth, it may develop in a more typical manner in WS than later emerging grouping abilities. This could be investigated by establishing the developmental time points of the emergence of grouping abilities in WS. However, this would be difficult to assess, as most individuals with WS are not diagnosed until late infancy at the earliest.

A relative strength in grouping ability in the closure task is also observed in WS. The ability to group by closure is consistent with the results of Wang et al., (1995) and Grice et al. (2003) (although note that the closure task here is strictly a similarity task with the topological properties of a closure task). Given the presence of topological properties in the closure task only, it is possible that the ability to group by topological properties is a relative strength in WS. Further investigation could compare grouping by closure to other forms of topological grouping properties, such as uniform connectedness. This is the principle that connected elements with uniform visual properties tend to be grouped together (Palmer & Rock, 1994). If elevated performance were also seen in WS on this task, this would support this suggestion of a relative strength in grouping by topological properties in WS.

The ability to group by orientation similarity is particularly poor in WS relative to the other forms of grouping measured. This could be due to a weak ability to discriminate between orientations. The Benton Line Orientation test (Benton et al., 1978) has been used to assess orientation discrimination ability in WS (Bellugi et al., 1988; Rossen et al., 1996; Wang et al., 1995). Results are generally reported to be at floor on this test. Thus, although a test that measured performance at the appropriate level would be more informative, these results appear to be consistent with the results of the present study, suggesting difficulty in processing orientation in WS.

The deviance in perceptual grouping ability demonstrated by the present study might also go some way to explain the global impairments experienced in image production (drawings or construction tasks) in WS. It would be interesting to systematically assess the nature of the global impairments in production in WS, according to the properties assessed in this study. This would determine the effect of deviant perceptual grouping abilities on their ability to produce images.

In summary, the present study showed an uneven profile of perceptual grouping abilities in WS. This is consistent with ERP evidence for unusual neural processing of perceptually grouped stimuli in WS (Grice et al., 2003). These results suggest that although individuals with WS are able to process images at the global level, this is not achieved in a typical manner.

References

Altmann, C. F., Bulthoff, H. H., & Kourtzi, Z. (2003). Perceptual organization of local elements into global shapes in the human visual cortex. *Current Biology, 13*, 342-349.

Atkinson, J., King, J., Bradick, O., Nokes, L., Anker, S., & Braddick, F. (1997). A specific deficit of dorsal stream function in Williams syndrome. *NeuroReport, 8*, 1919-1922.

Bellugi, U., Sabo, H., & Vaid, J. (1988). Spatial deficits in children with Williams Syndrome. In J. Stiles-Davis & U. Kritchevshy & U. Bellugi (Eds.), *Spatial Cognition: Brain Bases and Development* (pp. 273-297). Hillsdale, New Jersey: Lawrence Erlbaum.

Ben-Av, M. B., & Sagi, D. (1995). Perceptual grouping by similarity and proximity: Experimental results can be predicted by intensity autocorrelations. *Vision Research, 35,* 853-866.

Benton, A. L., Varney, N. R., & Hamsher, K. d. (1978). Visuospatial judgement: A clinical test. *Archives of Neurology, 35*, 364-367.

Bihrle, A., Bellugi, U., Delis, D., & Marks, S. (1989). Seeing either the Forest or the Trees: Dissociation in Visuospatial Processing. *Brain and Cognition, 11*, 37-49.

Bremner, G. (1994). *Infancy*. Oxford, England: Blackwell.

Braddick, O., O'Brien, J. M. D., Wattam-Bell, J., Atkinson, J., Hartley, T., & Turner, R. (2001). Brain areas sensitive to coherent visual motion. *Perception, 30*, 61-70.

Braddick, O., Atkinson, J., & Wattam-Bell, J. (2003). Normal and anomalous development of visual motion processing: motion coherence and 'dorsal stream vulnerability'. *Neuropsychologia, 41*, 1769-1783.

Chen, L. (1986). Topological perception- A possible dark cloud over computational theory. In Q. X. Sen (Ed.), *Essays on Cognitive sciences* (pp. 250-301). Shanghai: People's press of Shanghai.

Farran, E.K., Jarrold, C. & Gathercole, S.E. (2001). Block Design performance in the Williams syndrome phenotype: A problem with mental imagery? *Journal of Child Psychology and Psychiatry, 42,* 719-728.

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, 23*, 175-202.

Farroni, T., Valenza, E., & Simion, F. (2000). Configural processing at birth: Evidence for perceptual organisation. *Perception, 29*, 355-372.

Galaburda, A., Holinger, D. P., Bellugi, U., & Sherman, G. F. (2002). Williams syndrome: Neuronal size and neuronal-packing density in primary visual cortex. *Achives of Neurology, 59,* 1461-1467.

Galaburda, A. M., & Bellugi, U. (2000). Multi-level analysis of cortical neuroanatomy in Williams syndrome. *Journal of Cognitive Neuroscience*, *12*, 74-88.

Goodale, M.A. & Milner, A.D. *Sight Unseen: An Exploration of Conscious and Unconscious Vision.* Oxford: Oxford University Press, 2004.

Grice, S. J., de Haan, M., Halit, H., Johnson, M. H., Csibra, G., Grant, J., & Karmiloff-Smith, A. (2003). ERP Abnormalities of illusory contour perception in Williams syndrome. *NeuroReport*, *14*, 1773-1777

Hamsher, K. D. S. (1978). Stereopsis and the perception of anomalous contours. *Neuropsychologia*, *16*, 453-459.

Han, S., Humphreys, G. W., & Chen, L. (1999). Uniform connectedness and classical Gestalt principles of perceptual grouping. *Perception and Psychophysics, 61,* 661-674.

Han, S., Song, Y., Ding, Y., Yund, E. W., & Woods, D. L. (2001). Neural substrates for visual perceptual grouping in humans. *Psychophysiology, 38,* 926-935.

Kapadia, M. K., Westheimer, G., & Gilbert, C. D. (1998). Spatial distribution and dynamics of contextual interactions in cortical area V1. *Society for Neuroscience Abstracts*, 789.786.

Kanizsa, G. (1978). Subjective contours. *Scientific American, 234*, 48-52.

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

Klein, B. P., & Mervis, C. B. (1999). Cognitive strengths and weaknesses of 9- and 10- year-olds with Williams syndrome or Down Syndrome. *Developmental Neuropsychology, 16*, 177-196.

Kohler, W. (1929). *Gestalt Psychology*. New York: Horace Liveright.

Kourtzi, Z., Tolias, A. S., Altmann, C. F., Augath, M., & Logothetis, N. K. (2003). Integration of local features into global shapes: Monkey and human fMRI studies. *Neuron, 37,* 333-346.

Kurylo, D. D. (1997). Time course of perceptual grouping. *Perception and Psychophysics, 59*, 142-147.

Lenhoff, H. M., Wang, P. P., Greenberg, F., & Bellugi, U. (1997). Williams syndrome and the brain. *Scientific American, 277*, 42-47.

Milner, A. D., & Goodale, M. A. (1995). *The Visual Brain in Action*. Oxford Psychological Press.

Mooney, C. M. (1957). Age in the development of closure ability in children. *Canadian Journal of Psychology, 11*(219-310).

Palmer, S. E., & Rock, I. (1994). Rethinking perceptual organisation: The role of uniform connectedness. *Psychonomic Bulletin and Review, 1*, 29-55.

Pani, J. R., Mervis, C. B., & Robinson, B. F. (1999). Global spatial organization by individuals with Williams syndrome. *Psychological Science, 10*, 453-458.

Quinn, P. C., Bhatt, R., Brush, D., Grimes, A., & Sharpnack, H. (2002). Development of form similarity as a gestalt grouping principle. *Psychological Science, 13*, 320-328.

Quinn, P. C., Burke, S., & Rush, A. (1993). Part-whole perception in early infancy: Evidence for perceptual grouping produced by lightness similarity. *Infant Behavior and Development, 16*, 19-42.

Raven, J. C. (1993). *Coloured progressive matrices*. Oxford, UK: Information Press Ltd.

Reiss, A. L., Eliez, S., Schmitt, J. E., Straus, E., Lai, Z., Jones, W., & Bellugi, U. (2000). Neuroanatomy of Williams syndrome: A high-resolution MRI study. *Journal of Cognitive Neuroscience, 12: supplement*, 65-73.

Ross, W. D., Grossberg, S., & Mingolla, E. (2000). Visual cortical mechanisms of perceptual grouping: interacting layers, networks, columns, and maps. *Neural Networks, 13*, 571-588.

Rossen, R., Klima, E. S., Bellugi, U., Bihrle, A., & Jones, W. (1996). Interaction between language and cognition: evidence from Williams Syndrome. In J. Beitchman & N. Cohen & M. Konstantareas & R. Tannock (Eds.), *Language,* *Learning and Behaviour disorders: Developmental, Behavioral and Clinical Perspectives* (pp. 367-392). New York: Cambridge University Press.

Schmitt, J. E., Eliez, S., Bellugi, U., Galaburda, A., & Reiss, A. L. (2002). Increased gyrification in Williams syndrome: evidence using 3D MRI methods. *Developmental Medicine and Child Neurology, 44*, 292-295.

Udwin, O., & Yule, W. (1991). A Cognitive and Behavioural Phenotype in Williams Syndrome. *Journal of Clinical and Experimental Neuropsychology, 13*, 232-244.

Ungerleider, L. G., & Mishkin, M. (1982). Two cortical visual systems. In D. J. Ingle & M. A. Goodale & R. J. W. Mansfield (Eds.), *Analysis of visual behaviour* (pp. 549-586). Cambridge, MA: MIT Press.

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.

Wertheimer, M. (1923). Untersuchungen zur Lehre von der Gestalt: II. *Psychologische Forschung, 4,* 301-350 [Partial translation in W.D. Ellis (Ed.) (1950). *A sourcebook of Gestalt psychology* (pp. 1971-1981). New York: Humanities Press.].

Woliver, R. E., & Sacks, S. D. (1986). Intelligence and Primary Aptitudes: Test Design and Tests Available. In R. Cattell, B & R. C. Johnson (Eds.), *Functional Psychological Testing: Principles and Instruments* (pp. 166-188). New York: Brunner/ Mazel.

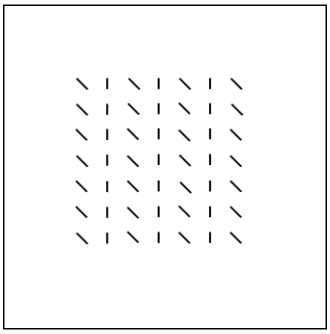
Table 1: Participant details

Group	CA	RCPM score	
	Mean (S.D.)	Mean (S.D.)	Range
WS	21;4 (11;8)	16.07 (5.60)	6-25
TD	5;2 (0;5)	16.33 (5.14)	9-26

Author Note

This study was supported by a grant to the author from the Reading Endowment Trust Fund. The author would like to thank those members of the WSF who have kindly participated in this study and the staff and students of Alfred Sutton Primary School for their co-operation with this work. Thanks also go to John Harris and David Milner (Editor) for useful discussion and comments on this manuscript. Correspondence concerning this article should be addressed to Emily Farran, School of Psychology, University of Reading, Earley Gate, Reading, RG6 6AL, UK. Electronic mail: E.K.Farran@reading.ac.uk

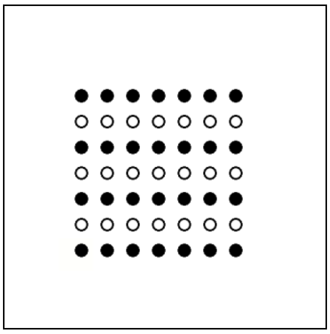
Figure 1: Similarity grouping types and difficulty levels



a. Category: Orientation

Grouping: Vertical

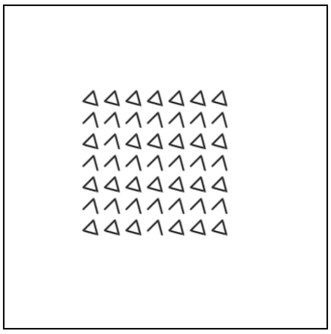
Level 1: 0 distracter elements



b. Category: luminance

Grouping: Horizontal

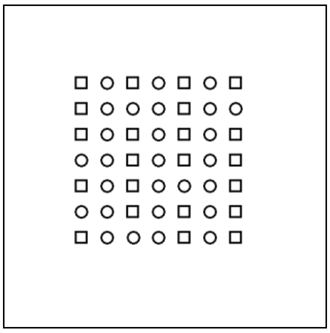
Level 1: 0 distracter element



c. Category: Closure

Grouping: Horizontal

Level 2: 1 to 3 distracters

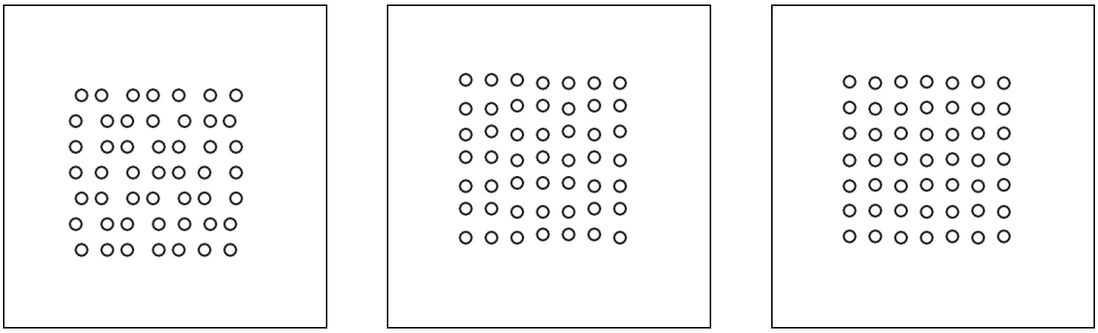


d. Category: Shape

Grouping: Vertical

Level 3: 4 to 6 distracters

Figure 2: Alignment: levels of difficulty



a. Grouping: Horizontal

Level 1

Misalignment: 9 pixels

b. Grouping: Vertical

Level 3

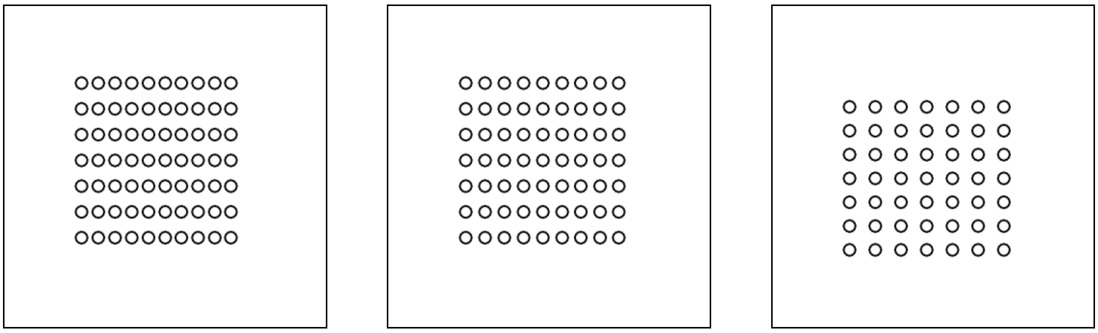
Misalignment: 5 pixels

c. Grouping: Vertical

Level 5

Misalignment: 3 pixels

Figure 3: Proximity: levels of difficulty



a. Grouping: Horizontal — Level 1: 5 pixels apart

b. Grouping: Horizontal — Level 3: 9 pixels apart

c. Grouping: Vertical — Level 5: 16 pixels apart

Figure 4: Similarity grouping threshold values (mean and standard error)

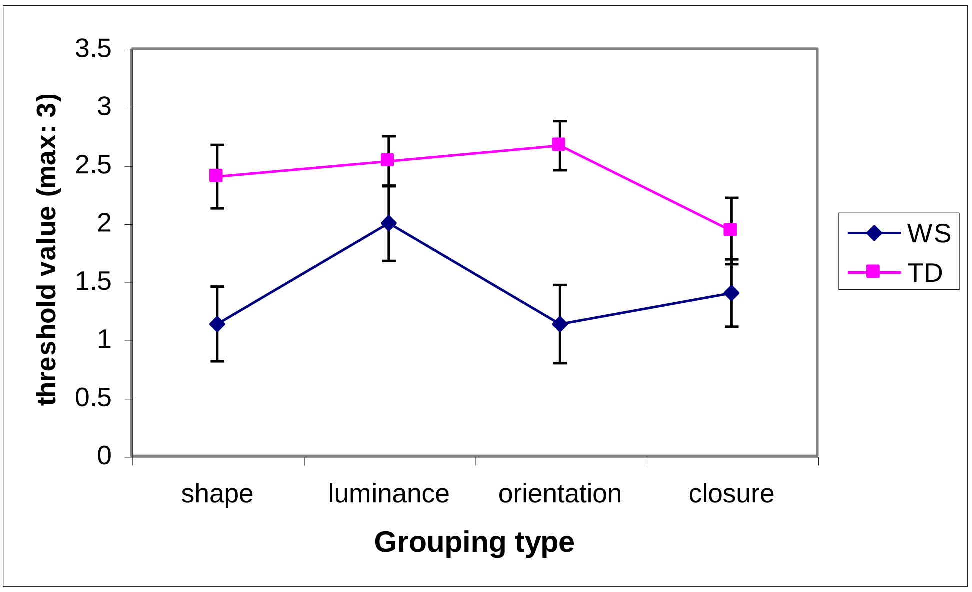


Figure 5: Spatial grouping threshold values (mean and standard error)

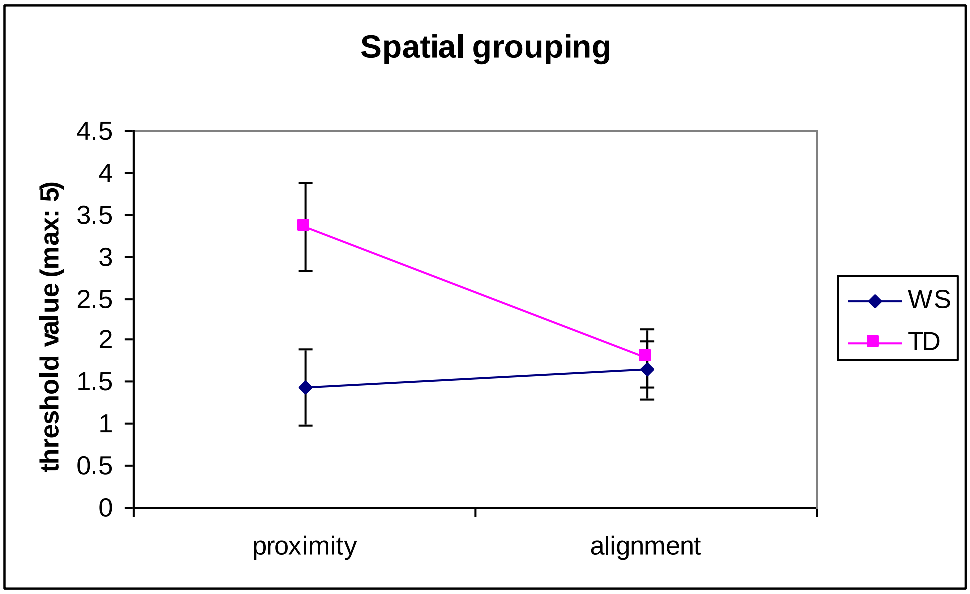


Figure 6: Profile of perceptual grouping abilities in WS (z-score mean and standard error)