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Evidence for Unusual Spatial Location Coding in Williams Syndrome: An Explanation for the Local Bias in Visuo-Spatial Construction Tasks?

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Abstract

Individuals with Williams syndrome (WS) display poor visuo-spatial cognition relative to verbal abilities. Furthermore, whilst perceptual abilities are delayed, visuo-spatial construction abilities are comparatively even weaker, and are characterised by a local bias. We investigated whether this differentiation in visuospatial abilities can be explained by a deficit in coding spatial location in WS. This can be measured by assessing participants' understanding of the spatial relations between objects within a visual scene. Coordinate and categorical spatial relations were investigated independently in four participant groups: 21 individuals with WS; 21 typically developing (TD) children matched for non-verbal ability; 20 typically developing controls of a lower non-verbal ability; and 21 adults. A third task measured understanding of visual colour relations. Results indicated first, that the comprehension of categorical and co-ordinate spatial relations is equally poor in WS. Second, that the comprehension of visual relations is also at an equivalent level to spatial relational understanding in this population. These results can explain the difference in performance on visuo-spatial perception and construction tasks in WS. In addition, both the WS and control groups displayed response biases in the spatial tasks. However, the direction of bias differed across the groups. This finding is explored in relation to current theories of spatial location coding.

Key words: Williams syndrome, Visuo-spatial cognition, Spatial location, Spatial relations.

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Introduction

Williams syndrome (WS) has an occurrence of approximately one in 20,000 births (Morris & Mervis, 1999) and is caused by a genetic deletion on the long arm of chromosome 7 (Nickerson, Greenberg, Keating, McCaskill, & Shaffer, 1995). The distinctive cognitive profile of this group is represented by significantly stronger verbal performance than non-verbal or visuo-spatial level of ability. It has been hypothesised that this comparative impairment of visuo-spatial functioning reflects a local processing bias in WS (e.g., Bellugi, Sabo, & Vaid, 1988), i.e., that an individual has a preference for processing the parts or the details of a visual array, over the whole or global image. However, recent research into the visuo-spatial cognition of individuals with WS has begun to question this assumption.

A local processing bias is often inferred from the pattern of performance of individuals with WS on the Block Design task (Wechsler, 1981) and in the drawing version of the Navon hierarchical processing task (Navon, 1977). The Block Design task requires the participant to assemble a number of blocks so that the pattern made by the upper faces of these blocks resembles a model image. The drawing version of the Navon task requires the individual to reproduce a hierarchical figure, for example, a number of letter As, arranged in the shape of a letter D, which represent local and global levels respectively. Participants with WS are reported to focus at the local level, i.e., the individual blocks in the Block Design task (e.g. Bellugi et al., 1988), and the local level letters in the Navon task (e.g., Bihrle, Bellugi, Delis, & Marks, 1989; Rossen, Klima, Bellugi, Bihrle, & Jones, 1996). This is at the expense of

organising these local level stimuli correctly to resemble the global form of the presented image.

At first blush, it does not seem unreasonable to consider that the broken configurations, offered as solutions by individuals with WS, merely indicate a delay in development. Indeed, young children often make configural errors on such tasks (Akshoomoff & Stiles, 1996; Kramer, Kaplan, Share & Huckeba, 1999). Furthermore, Kramer et al. (1999) demonstrated a correlation between local processing preferences at the perceptual level and configural errors on construction tasks, thus a local bias in WS seems an appropriate hypothesis. However, there are two crucial differences between WS performance and that of typically developing young children. First, individuals with WS are unable to correct their configural errors (Hoffman, Landau & Pagani, 2003), whilst the majority of young children correct their configural errors before offering their final solution (Kramer et al., 1999). The second, discussed below, relates to perceptual processing in WS.

Farran, Jarrold, and Gathercole (2003) employed both the drawing version of the Navon Task (as described above), and perceptual versions of the task in which the participants had to attend to the local and global levels of the stimuli either sequentially or simultaneously. Results demonstrated that individuals with WS were perceptually sensitive to both local and global levels of processing to the same extent as typically developing (TD) controls matched by non-verbal ability. Despite these results in the perceptual tasks, in the drawing task, as with previous studies, the WS group represented the local elements in unstructured formats, and thus a significant difference was observed between the groups in this version of the task. These results challenge a local processing bias hypothesis and stand in contrast to the patterns observed in typical development, as, perceptually, individuals with WS do not appear

to show a bias to process locally. It is only once an output is required, in this case drawing, that local elements become more salient than the global form.

Further evidence against a local processing bias at the perceptual level comes from performance on the segmented version of the Block Design task. In this task, the individual blocks of the model image are slightly separated, which has the effect of making each local element more salient, thus the magnitude of a local processing preference in WS at perception can be investigated (cf. Shah & Frith, 1993). Mervis, Morris, Bertrand, and Robinson (1999) reported a facilitation effect of segmentation in a group of individuals with WS. Furthermore, Farran, Jarrold, and Gathercole (2001) demonstrated that the facilitation effect of segmentation in the performance of individuals with WS did not differ significantly from that observed in a group of TD controls matched by non-verbal ability. This suggests that perception in WS is no more influenced by local information than in the typically developing population, and thus these results are not consistent with a local processing bias account. Importantly, as above (Farran et al., 2003), this result contrasts with the pattern of construction abilities in the Block Design task in WS, where a local bias is apparent.

To fully account for these data, a theory is required which can explain how global impairments become apparent in the drawing or construction of an image despite apparently typical perceptual processing preferences of that same image. One such account could relate to the procedural difference between these tasks. In order to complete drawing and construction tasks accurately, the individual must first deconstruct the image into its local elements, and then reconstruct the image to create their reproduction. Knowledge of the spatial location of the parts of the image (e.g., the small letters or blocks) is crucial to reproducing the image. This requires encoding the spatial relationships between the elements within the global figure. Without

encoding an element's location in relation to other elements, the image risks being reproduced in a manner in which each element is placed haphazardly, i.e., with no global cohesion. This appears to be the pattern seen in the reproductions of individuals with WS.

In contrast to the demands of production tasks, coding location is less important to the completion of perceptual tasks. In these tasks, the spatial relations between the parts of an image serve only to aid object recognition. As such, an ability to encode spatial relations is not vital to successful task completion in perceptual tasks. If the coding of spatial relations is problematic in WS this could explain the dissociation in performance between perceptual and production tasks. It could also explain the distinct, piecemeal pattern of the reproductions given by individuals with WS. Thus, we hypothesise that individuals with WS may have a poor comprehension of spatial location due to an impaired or deviant ability to encode spatial relations.

Hoffman et al. (2003) offer some support for the present hypothesis. In order to successfully place each element in the correct location when reproducing an image, an individual must continuously monitor the spatial relations between elements by alternately fixating on the model image and the part-finished solution (Ballard, Hayhoe, Pook, & Rao, 1997; Lagers-van Haselen, van der Steen, & Frens, 2000). Hoffman et al. (2003) report that in a block construction task, individuals with WS made fewer saccades between the model image and the part-finished solutions than TD controls of the same IQ. If, as hypothesised, individuals with WS are impaired at encoding spatial relations, compared to the typical population, they may find it less beneficial to monitor these spatial relations. Thus, the reduction in monitoring in WS could reflect a poor representation of space.

Phillips, Jarrold, Baddeley, Grant, and Karmiloff-Smith (2004) demonstrated an impairment in the understanding of spatial language through words such as 'above' and 'below', 'in' and 'on', 'in front' and 'behind' in WS, in comparison to controls matched by vocabulary level. Specifically, this result points towards a possible problem in coding spatial relations when linguistic terms are necessary for referencing spatial categories (see Hayward & Tarr, 1995). More generally, this deficit in spatial language in WS may be indicative of a similarly weak non-linguistic ability at encoding spatial categories (see e.g., Brown, 1973; Clark, 1973; Bowerman, 1996).

The encoding of space, in the typical population, has been described by Huttenlocher, Hedges, and Duncan. (1991) in their Category Adjustment Model (CAM). The CAM makes a distinction between 'categories' and 'particulars' for coding location. These two terms are essentially comparable to Kosslyn and Koenig's (1992) 'categorical spatial relations' and 'coordinate spatial relations' respectively. Particulars/ coordinate spatial relations refer to the encoding of fine grain information. This information is used to refer to precise locations, or specific distances between elements, e.g., that the chair is a certain distance from the table. Categories are regions of space which cover a range of particular values (Huttenlocher et al., 1991).

The present study, uses tasks adapted from Kosslyn et al. (1989) and Koenig, Reiss, and Kosslyn. (1990) to measure the encoding of categorical and coordinate spatial relations in WS. Kosslyn et al. (1989) employed two computer tasks in which a horizontal line remained in the centre of the presentation screen, and a small square appeared either above or below the line, at one of six distances from the line. In the categorical task, the participant had to indicate whether the square was 'above' or 'below' the line. In the co-ordinate spatial relations task, the individual had to indicate whether the square was within 3mm of the horizontal line, or further than 3mm from

the line. Koenig et al. (1990) used these tasks with children. The authors explained the rules by disguising the task as a baseball game with the square as the ball, and the line as the bat.

An adaptation of Koenig et al.'s (1990) version of the tasks was employed here. A test of visual colour relations was also included in order to provide a marker of the comprehension of visual relations in WS relative to that of spatial relations. In the colour task, the individual was asked to assess category membership, blue or green, of a colour patch that varied systematically between the hues of prototypic green and blue colour patches.

We assessed performance not only for absolute accuracy as in the study by Koenig et al. (1990), but also in terms of patterns of performance as this gives insight into the strategies employed to adjust inexact representations. This can be related to Huttenlocher et al.'s (1991) CAM, which describes how, in typical development, inexact representations are adjusted using category information; category boundaries (the value at the endpoint of a category) and prototypes (a category exemplar falling in the centre of a category). They also note that category boundaries or prototypes can also be also inexact, which can lead to less effective adjustments. Visual information can also be coded using the CAM (e.g., a specific orange colour is a particular of the category 'orange', the category 'orange' has boundaries with red and yellow, and a prototypical value).

The levels and patterns of performance were observed for the two spatial tasks and the visual task. It was hypothesised that individuals with WS would show differential performance to that observed in non-verbal matched typical developing controls. Post hoc comparisons to CA matched controls, and young typically developing controls (matched by level of ability) explored the patterns of performance

of individuals with WS and whether these patterns fit the typical developmental trajectory.

Method

Participants

Four participant groups were employed; Twenty-one individuals with WS, 21 typically developing controls matched individually by ability on the Ravens Coloured Progressive Matrices (RCPM; Raven, 1993), 20 young controls of a lower level of RCPM performance, and 21 chronological matched TD adults. The RCPM is a non-verbal perceptual task in which the participant must decide which of 6 pieces matches the pattern or follows the correct sequence shown in a presented image. This task was chosen to measure nonverbal ability, as it is an accepted measure of fluid intelligence (Woliver & Sacks, 1986). As such, performance can be assumed to be an appropriate representation of general nonverbal ability.

The WS group were originally recruited through the Williams Syndrome Foundation for a previous experiment, and were familiar with experimental testing procedures. A diagnostic "fluorescence in situ hybridisation" test (FISH) had been administered to 6 of these individuals, confirming deletion of the elastin gene on chromosome 7q11.23 in each of these cases. This gene is deleted in approximately 95% of individuals with WS (Lenhoff, Wang, Greenberg, & Bellugi, 1997). The remaining 15 participants were diagnosed before the FISH test became available as a diagnostic tool. Diagnosis was made by medical practitioners on the basis of phenotypic characteristics; all individuals had the recognisable 'elfin face' and social personality; medical characteristics such as heart defects and the presence of hypercalcaemia in infancy were common in the group. Additionally, these individuals

showed a cognitive profile (see Mervis, 1999) which was characteristic of WS, by exhibiting, for example, very poor performance on block construction tasks, and relatively superior verbal comprehension (see Farran, Jarrold & Gathercole, 1999).

The typically developing (TD) children were recruited from local main stream schools in Bristol (matched TD group) and Reading (young TD group), and the adult typically developing group were students at the University of Reading. Two of the participant groups, the young TD and adult groups, were recruited post hoc in order to facilitate the interpretation of the results of the categorical and coordinate spatial relations tasks. The adult group were matched to the WS group, as a group, by chronological age (CA). The young TD group were approximately matched to the WS group for level of ability across the spatial relations tasks. Data from these two groups are reported for the two spatial tasks only. Participant details can be seen in Table 1.

Design and Procedure

Three computerised tasks were administered measuring categorical spatial relations, co-ordinate spatial relation, and visual relations respectively. In each task, the participant was presented with instructions both on the computer screen and verbally by the experimenter.

Categorical relations task

In this task, the individual was told that a man would appear on the screen holding a baseball bat. The man was positioned at the left side of the screen standing vertically, holding a bat horizontally along the centre of the screen. The individual was told that a ball would appear on the screen which the man had already hit either up or down. If the ball had been hit up, it appeared above the bat. If the man had hit the ball down, the ball would appear below the bat. It was explained that participants

were to decide whether the ball had been hit up (above) or down (below) by pressing response keys. The 'P' key and 'W' keys corresponded to the up/above and down/below responses respectively. The experimenter ensured that the participant understood the two categories using both verbal labels (above, up, below, down) and by pointing to the areas on the screen. There were 12 practise trials and 48 experimental trials. All participants carried out one block of 12 practise trials, as the experimenter was confident that the procedure had been understood. The ball could appear in one of 12 positions, 6 positions above the bat and 6 positions below the bat/ This is illustrated in Figure 1a, and described below in terms of pixel size and degrees of visual angle. The pixel setting was set at 72 pixels/inch (28.35pixels/cm) with a viewing distance of 50cm. Pilot testing was conducted in order to equate levels of difficulty between the spatial relations tasks. Thus in this task, the bat was shorter than in the co-ordinate task below, and the balls more closely spaced. The bat was 24 pixels (0.49°) wide at the widest point, and had a length of 138 pixels (2.79°). The balls were 10 pixels (0.20°) in diameter, with a horizontal distance of 55 (1.11°) pixels between the tip of the bat and the centre of the ball. The vertical positions of the centres of the balls were equally spaced at 5 pixels (0.10°) apart, with a 5 pixel (0.10°) distance between the centre of the first ball above or below the bat, and the vertical centre of the bat. The task took approximately 5 minutes to complete.

Co-ordinate relations task

In this task, the same man, again holding a horizontal bat appeared on the left side of the screen. The individual was told that the ball was approaching the man and he could only hit it if it was near enough to his bat. This was described as being 'in', as in a game of rounders or baseball. Balls that were further away were described as being 'out'. An example appeared on the screen in which all 12 of the positions that

the ball would appear were shown simultaneously, as shown in Figure 1b. This displayed the 6 balls that were close to the bat ('in'), 3 above and 3 below the bat, and 6 balls that were further away from the man's bat ('out'). Two horizontal lines, one above and one below the bat, indicated the division between near/'in' and far/'out'. As above, the experimenter ensured that the participant understood the two categories using both verbal labels (near, in, far, out) and by pointing to the areas on the screen. There were 12 practise trials, one for each ball position, in which the horizontal lines remained on the screen for the participant to learn the correct co-ordinates which indicated whether the man could hit the ball (near/ 'in') or could not hit the ball (far/ 'out'). Again, for all participants, the experimenter was confident that they understood the procedure after one block of 12 practise trials. Thus the experimental trials followed after one practise block. The 48 experimental trials did not feature the horizontal dividing lines, so that the individual had to judge co-ordinate distance. In all trials, correct responses on each trial were required in order for the programme to progress through each trial. Participants were instructed to press the 'X' key and the ',' key for 'in' and 'out' responses respectively. As above, the pixel setting was set at 72 pixels/inch (28.35pixels/cm). The bat was 24 pixels (0.49°) wide at the widest point, but in this task was 207 pixels (4.18°) in length. The balls were 10 pixels (0.20°) in diameter, and remained in the same horizontal position as in the task above, but due to the longer bat length, the balls were aligned vertically with the widest part of the bat. The positions of the balls were equally spaced 20 pixels (0.40°) apart with a 20 pixel (0.40°) distance between the centre of the first ball above or below the bat, and the vertical centre of the bat. The dividing lines between the 'in' trials and the 'out 'trials were 70 pixels (1.41°) in horizontal length when shown in the practice trials, and were positioned directly between the 3rd and 4th balls above and below the

bat, at a vertical distance of 10 pixels (0.20°) from the centre of each ball. The task took approximately 5 minutes to complete.

Colour relations task

This task was included as a visual comparison to the spatial tasks and was completed by the WS and matched TD control group only. The computer displayed three coloured squares with a black border, on a white background. 'Paint shop pro' software was used to adjust the hues of these colours. The colour of the square on the left and of the square on right was kept constant. The left square was set at maximum hue value for blue, with no green or red input, i.e. a pure blue (hue: 160, R:0, G:0, B:255), whilst the square on the right was set at maximum hue value for green, with no blue or red input, i.e. a pure green (hue: 80, R:0, G:255, B:0). Participants were instructed that the colour of the central square was a mixture between blue and green, and that they were to decide if it was more like the blue square or more like the green square by pressing 'S' and '=' respectively. The hue of the central square was adjusted by systematically decreasing the quantity of blue colour hue as the amount of green colour hue increased. There was never any input from red. This created 8 trials in which the colour of the central square spanned the blue-green spectrum. In 4 of these trials, the central square had a hue more similar to the pure blue square than the green square (stimulus 1: hue: 138, R:0, G:140, B:255; stimulus 2: hue: 135, R:0, G:159, B:255; stimulus 3: hue: 132, R:0, G:179, B:255; stimulus 4: hue: 129, R:0, G:198, B:255) and 4 trials in which the central square was of a hue more similar to the pure green square than the blue square (stimulus5: hue: 111, R:0, G:255, B:198; stimulus 6: hue: 108, R:0, G:255, B:179; stimulus 7: hue: 105, R:0, G:255, B:159; stimulus 4: hue: 102, R:0, G:255, B:140). The task included 8 practise trials, one of each trial type, and 32 experimental trials, 4 of each trial type presented in random

order. Each trial followed after an individual had made a response, independent of whether the response was correct or not. This was so that the individual could not learn to recognise the colours and the appropriate response that corresponded to that colour. Total experiment running time was about 5 minutes.

Results

Data for each task were analysed separately. These results are presented first, followed by a comparison of the level of performance across the three tasks. Separate ANOVAs of the number of correct responses are reported for each task in terms of within- and between-participant effects and within-participant contrasts. Within participant contrasts are reported for factors where the significance of the linear trend of the levels within the factor is of interest, i.e., in the effect of the linear increase in spatial (categorical and co-ordinate tasks) or visual (colour task) distance of the stimulus from the midpoint, on the dependent variables. Reaction time (RT) data were similarly analysed. There were 17 (1.7%) empty cells in the categorical task (WS: 7 cells, matched TD: 1 cell, young TD: 9 cells) and 4 (0.8%) empty cells in the coordinate task (young TD: 4 cells) due to floor effects in responses to certain trials. To account for this, the average value for that participant group and at that trial type replaced the missing values. In the colour relations task, it was not possible to analyse RT due to too many missing values for the 'green' responses.

Categorical spatial relations

The number of correct responses in the categorical spatial relations task are illustrated in figure 2a. These data were analysed by a three factor repeated measures ANOVA, with category (2 levels; above and below), and position (position of the ball from the bat, 6 levels, labelled position 1 to 6 with increasing distance from the bat) as

within participant factors). The between participant factor was group (4 levels: WS, matched TD, adult, young TD). Analysis showed a significant main effect of group, F(3,79)=14.65, p<.001, partial $\eta^2 = .36$. Post hoc Tukey comparisons revealed that this was due to significantly higher performance from the adult group compared to the WS and young TD group (p<.001 for both), and significantly higher performance from the matched TD controls than the young TD group (p=.002). The main effect of position was also significant, reported as a linear trend, F(1, 79)=15.46, p<.001, partial $\eta^2 = .16$, due to an increase in accuracy with distance from the bat. The main effect of category was not significant, F(1, 79) = 1.71, p=.19, partial $\eta^2 = .02$. There was a significant interaction between position and group, F(15, 395) = 3.31, p < .001, partial $\eta^2 = .11$. This reflected a linear effect of position in the WS and matched TD groups, whilst the adults were at ceiling on many trials, and the variability of the young TD group did not appear systematic (WS, p=.004; matched TD, p=.001; young TD, p=.54; adults, p=.054). There was also a significant interaction between category and group, F(3, 79)=3.37, p=.02, partial $\eta^2=.11$. Post hoc Tukey analysis demonstrated that this was due to differential effects of group in the two categories: for above responses the WS group were most similar to the matched TD controls (WS and matched TD, p=.85, WS and adults, p=.001, WS and young TD, p=.06); whilst for below responses WS performance most resembled the level of the young TD group (WS and matched TD, p=.01, WS and adults, p<.001, WS and young TD, p=1.00). The remaining interactions were not significant (category by position: F(5,395)=1.09, p=.37, partial $\eta^2 = .01$; category by position by group, F < 1).

Figure 2a shows that this significant interaction may be the result of different response biases from the WS group, compared to the control groups. The minimum number of correct responses occurred at position 1 below the bat in the WS group, but

occurred at position 1 above the bat in the control groups. To investigate the possibility of differential group biases, a second 3-way, repeated measures ANOVA was conducted in which the data from the WS group, as it is displayed in Figure 2a, was shifted to the left so that the lowest data points were aligned across the four groups. This central point was re-labelled as position 0 (see Figure 2b). Thus, there were 5 positions on each side of position zero in which ball position was increasingly further away from the bat. For this analysis, the responses for positions one to five on each side of position zero were compared. There were 2 within participant factors; category (2 levels; above and below); and position (5 levels), and one between participant factor of group (4 levels; WS, matched TD, adult, young TD). This further analysis revealed that by shifting the data points in this way, the interactions between group and category and between group and position were no longer significant (group by category, F<1; group by position, F(12, 316)=1.46, p=.14). One can infer from this that the previous significant interactions in the original ANOVA above, were due to differences in response bias between the groups, rather than fundamental differences in the ability to respond to categories.

Slope values were calculated for each participant to investigate this response bias further. These were based on the slopes between a 'central' point (position 1 above or below the bat) and the adjacent data point on either side. The two slopes on either side of the central point were compared using paired t-tests. A non-significant t-test indicates that the slope values are symmetrical and thus that the point allocated as the 'central' point is a true reflection of the lowest level of performance of the group. Results revealed that either data point could be labelled the 'central' point for the young TD controls (p>.05 for both). However, this is not surprising as this group did not show a significant effect of position. For the remaining three groups, results

revealed that the position 1 below the bat was a suitable central point for the WS group (WS, t(20)=-.31, p=.76) but not the matched TD group (t(20)=-3.01, p=.01) or the adult group (t(20)=2.50, p=.72). In contrast position 1 above the bat was a suitable central point for the matched TD group (t(20)=-.384, p=.71) and the adult group (t(20)=-.37, t=.72), but not the WS group (t(20)=2.42, t=.03). This analysis, again, supports the suggestion that the WS group have a differential response bias to the control groups. The pattern of responses of the WS group indicated that they perceived the division between the 'above' and 'below' categories to be below the true category division point. In contrast, the three typically developing groups showed a response bias in the opposite direction, making the most errors at a higher position than the correct division point.

As with the correct response data above, a 3 way repeated measure ANOVA was carried out on RT, also with two within participant factors; category (2 levels; above and below); and position (6 levels), and one between participant factor of group (4 levels; WS and matched TD, adult, young TD). The main effect of group was significant, F(3, 79)=12.04, p<.001, partial η^2 =.31. Tukey post hoc comparisons revealed that this was due to significantly shorter RTs in the adult group compared to the remaining three participant groups only (p<.001 for all). The WS, matched TD and young TD groups had comparable RTs (p>.05). There was a significant main effect of position, reported here in terms of within-participant linear contrasts, F(1, 79)=9.18, p=.003, partial η^2 =.10, which indicates that RT increased linearly as distance from the bat decreased. The main effect of category was not significant (category: F(1, 79)=1.15, p=.29, partial η^2 =.01). There were no significant interactions (category by group: F(1, 79)=1.13, p=.34, partial η^2 =.04; position by

group: F(15, 395)=1.19, p=.28, partial $\eta^2 = .04$; category by position: F(5, 395)=1.05, p=.39, partial $\eta^2 = .01$; category by position by group: F<1).

Co-ordinate relations task

Analysis of the number of correct responses was performed by collapsing the data at the position of the bat (the 'midpoint') into six data points, 3 'in' and 3 'out' responses. A 3 factor ANOVA was employed, with 2 within participant factors. These were; co-ordinate classification (2 levels; 'in' and 'out') and position (3 levels of ball position, labelled positions 1 to 3 with increasing distance from the co-ordinates of the 'in'/'out' division point). There was one between participant factor of group (4 levels: WS, matched TD, adult, young TD). The results of this analysis, as illustrated in figure 3a, showed a significant main effect of group, F(3, 79)=26.25, p<.001, partial η^2 = .50. Tukey post hoc analysis revealed that the young TD group performed at a significantly lower level than the other participant groups (p<.001 for all). The performance of the WS, matched TD, and adult groups did not differ significantly (p>.05 for all). There was a significant main effect of position, reported as a linear contrast, F(1, 79)=169.46, p<.001, partial $\eta^2=.68$, thus indicating that the number of correct responses decreased linearly with decreasing distance from the co-ordinates of the dividing point. The main effect of co-ordinate classification was also significant, F(1, 79) = 22.26, p < .001, partial $\eta^2 = .22$. This is accounted for by overall poorer performance on 'out' trials than 'in' trials. There was also a significant interaction between co-ordinate classification and group, F(3, 79)=4.35, p=.007, partial $\eta^2=.14$. This resulted from a difference in the effect of group for 'in' and 'out' responses (Tukey post hoc comparisons): For 'out' responses, WS performance was significantly higher than the young TD group (p<.001), but at a comparable level to

the adult (p=.98) and matched TD groups (p=.85). For 'in' responses, WS performance remained significantly higher than the young TD group (p=.04), but was significantly lower than the adult (p=.001) and matched TD groups (p=.009). The interaction between position and group was also significant, F(6, 158)=3.06, p=.007, partial $\eta^2 = .10$. All four groups demonstrated a significant effect of position (p<.05). Thus, the source of the interaction was due to differential group effects at each position. Tukey analysis revealed that WS performance was more similar to adult and matched TD performance at the position nearest the bat (position 1), (WS and matched TD, p=.53; WS and adults, p=.85; WS and young TD, p=.001), than the middle position (position 2) (WS and matched TD, p=.09; WS and adults, p=.007; WS and young TD, p<.001) and the position furthest from the bat (position 3) (WS and matched TD, p=.07; WS and adults, p=.06; WS and young TD, p<.001). The interaction between coordinate classification and position was also significant. This was due to differences in the effect of coordinate classification with position: position 1, F(1, 79)=15.83, p<.001, partial $\eta^2=.17$; position 2, F(1, 79)=3.77, p=.056, partial $\eta^2 = .05$; position 3, F(1, 79) = 14.70, p < .001, partial $\eta^2 = .16$. There was also a significant 3 way interaction of co-ordinate classification by position by group, F(6,158)=5.19, p=.001, partial $\eta^2=.17$. Further analysis revealed that the source of this interaction was a significant interaction between group and co-ordinate classification at position 1, F(3, 79)=6.60, p<.001, partial $\eta^2=.20$ and position 3, F(3, 79)=2.89, p=.04, partial $\eta^2 = .10$, but not at position 2 (F(3,79)=1.79, p=.16, partial $\eta^2 = .06$). The interaction at position 1 was due poorer performance in the adult and matched TD groups on the 'out' trials than the 'in' trials (adults: t(20)=4.57, p<.001; matched TD: t(20)=3.30, p=.004). In contrast, the WS and young TD groups showed no significant difference between these two data points (WS: t(20)=-1.24, p=.23; young TD: t(19)=

.95, p=.35). At position 3, all groups showed poorer 'out' performance than 'in' performance, except for the young TD group (WS, matched TD, adults, p<.05 for all; young TD, p=.16).

Figure 3a enables one to inspect the data points at which the most errors were made in each group. The pattern of response accuracy suggests (as in the categorical task above) that the WS and control groups had different response biases. Thus, for further analysis, the data for the groups were shifted so that the data point with the most errors, for each respective group, lay on top of one another. The most errors were made in position 1 of the 'in' responses for the WS group, and position 1 of the 'out' responses for the matched TD, adult and young TD groups. These points were renamed as position 0. Four data points remained, 2 on each side of zero representing 'in' and 'out' responses respectively. These are shown in Figure 3b. A second ANOVA was carried out on these re-coded data, again with 3 factors. The within participant factors were co-ordinate classification (2 levels; 'in' and 'out'), and position (2 levels), and the between group factor was group (4 levels; WS, matched TD, adult, young TD). This analysis showed that only the main effect of group and of position remained significant. This supports the suggestion that the main effect of coordinate classification and the 2-way and 3-way interactions in the original ANOVA were due to the differential response biases of each group.

Slope analysis was carried out to further investigate these response biases. As with the categorical task, slopes were based on the slope between a 'central' point (position 1 for 'in' or 'out' responses) and the adjacent data point on either side of this central position. The two slopes on either side of the central point were then compared using paired t-tests. Results showed that the 'in' position 1 was the most appropriate central point for the WS group (WS: t(20)=1.14, p=.27; matched TD: t(20)=3.77,

p=.001; adults: t(20) = 7.38, p<.001; young TD: t(19)=3.07, p=.006), whilst the 'out' position 1 was the putative central point for the three typically developing groups (WS: t(20)=-2.75, p=.012; matched TD: t(20)=-0.37, p=.71; adults: t(20)=-1.096, p=.29; young TD: t(19)=0.14, p=.89). This supports the analysis above, and suggests that the WS group were biased to perceive the 'in'/'out' division point as nearer the centre of the image, whilst the three typically developing groups showed the opposite bias, by perceiving the division point as further towards the outer edge of the image than the true division point.

Analysis of RT data was also performed by collapsing the data at the position of the bat (the 'midpoint') into six data points, 3 'in' and 3 'out' responses. Once again, a 3 factor ANOVA was performed with 2 within participant factors; coordinate classification (2 levels; 'in' and 'out') and position (3 levels); and one between-participant factor of group (4 levels: WS, matched TD, adult, young TD). Results showed a significant main effect of group, F(3, 79)=13.09, p<.001, partial η^2 =.33. Tukey analysis demonstrated that adult performance was faster all other groups (p<.05). RTs amongst the WS, matched TD and young TD groups did not differ significantly (p>.05). The significant main effect of position is reported in terms of linear contrasts; F(1, 79)=5.42, p=.023, partial $\eta^2=.06$, and reflects faster RTs with increasing distance from the 'in'/'out' dividing line. The main effect of co-ordinate classification was not significant (F<1). There were no significant interactions (coordinate classification by group: F(3, 79)=1.21, p=.31, partial $\eta^2 < .04$; position by group: F(6, 158)=2.12, p=.054, partial $\eta^2=.08$; co-ordinate classification by position: F(2, 158)=2.08, p=.13, partial $\eta^2=.03$; co-ordinate classification by position by group: F(6, 158)=1.75, p=.11, partial $\eta^2=.06$).

Colour task

The WS and matched TD controls only, took part in this task. Correct responses were analysed by a 3 way ANOVA. There were 2 within participant factors: colour (2 levels: blue and green); and hue (4 levels of increasing difference in hue between the pure colour, blue or green, to a central blue-green colour) and one between participant factor of group (2 levels: WS, matched TD controls). Analysis revealed a main effect of group, F(1, 40) = 14.61, p < .001, partial $\eta^2 = .27$, with the WS group scoring less accurately than the matched TD controls. There was a main effect of colour, F(1, 40)=27.25, p<.001, partial $n^2=.41$ due to poorer performance on the green colour trials, than the blue colour trials. This was an unexpected result and was analysed further as described below. There was also a main effect of hue as indicated by a significant linear contrast, F(1, 40)=41.55, p<.001, partial $\eta^2=.51$. There was also a significant interaction which occurred between colour and hue, reported as linear contrasts, F(1, 40)=7.37, p=.01, partial $\eta^2=.15$. The source of this interaction was a larger linear trend as hue changed across trials in the green trials, F(1,40)=27.64, p<.001, partial η^2 = .41, than in the blue trials, F(1, 40)=6.85, p=.01, partial $\eta^2 = .15$. There were no other significant interactions (colour by group: F(1, 40)=3.55, p=.07, partial η^2 = .08; hue by group: F(3, 120)=1.89, p=.14, partial η^2 = .05; colour by hue by group: F(3, 120)=1.46, p=.23, partial η^2 = .04). These data are illustrated in Figure 4a below.

The main effect of colour was a counter-intuitive result. Both groups of individuals showed a tendency to give a 'blue' response rather than a 'green' response, resulting in reduced accuracy in the green trials. The task was designed, using colour patches from the 'paintshoppro' software package so that the 'green' stimuli and the 'blue' stimuli were of equal differences in hue from the pure green and

the pure blue respectively i.e. it was thought that hue was symmetrical about the midpoint. The hue of each colour patch was further investigation using a chromometre. This measures the x and y chromaticity co-ordinates of a colour, which enabled a comparison to be made between the colour hues of the colour patches by plotting these x and y co-ordinates in 2D space. The chromometre revealed that the x, y co-ordinate chromaticity readings were not symmetrical about the midpoint (see figure 4b). The 'green' stimuli were closer to the midpoint, and further away from the pure green colour, than the equivalent 'blue' stimuli. Put simply, the 'blue' stimuli were more blue, than the 'green' stimuli were green. This can explain the imbalance in the pattern of results and the resulting main effect of colour.

Comparison across tasks

Performance across all three tasks was compared: first, to compare performance between the two spatial tasks relative to typical development. Second, to compare performance on the spatial tasks, to that on the visual task, the control task. Two of the four participant groups participated in all three tasks, the WS group and the TD matched control group. Thus, in order to compare performance, the mean number of correct responses of the WS group for each task were converted into z-scores based on the distribution of performance of the matched TD control group. These scores indicate the extent to which the WS group were performing differently or similarly to the matched TD group on each task and thus allows performance across the three tasks to be compared. Z-scores were compared using a one-factor ANOVA with three levels of task: categorical, co-ordinate, and colour. There was no main effect of task, F(2, 40)=1.55, p=.23, partial $\eta^2=.07$ indicating that the WS group

were similarly impaired across tasks, relative to the performance of the matched TD control group.

Discussion

This study compared the performance of individuals with WS to that of three other participant groups for the spatial tasks and one participant group for the visual task. Comparisons were to determine, first whether the level of performance of individuals with WS differed from their general level of visuo-spatial ability, and second whether the pattern of performance of individuals with WS was observed in the typically developing population. The matched TD controls (matched by general level of visuo-spatial cognition) participated in all three tasks. The adult group and young TD group were added post-hoc to further investigate the typical trajectory of patterns of performance on the spatial tasks.

The *level* of performance shown by individuals with WS was consistently lower than that of the matched TD group, although only significantly so for the colour task. A comparison across tasks indicated that WS performance across the three tasks did not differ significantly, relative to the performance of the matched TD controls. Thus, it seems that individuals with WS can make relational comparisons between objects for classification. The level of this ability is at a level similar to or below their general level of visuo-spatial cognition ability and appears to be independent of whether the comparisons are based on spatial or on visual relations.

More precise levels of performance can be ascertained on the spatial tasks.

Comparisons with the adult and young TD groups demonstrated that, when encoding categorical spatial relations, the performance of individuals with WS does not differ from the level reached by a typically developing child aged between four-and-a-half (young TD group) and six (matched TD group), but is lower than the adult level of

ability. When encoding coordinate spatial relations, individuals with WS perform at a significantly higher level than a typically developing four-year-old, but did not differ from the level of a typically developing six-year-old (matched TD controls). The performance of individuals with WS was also not different to that of typically developing adults, however, the true adult level of performance on the easier trials appears to be masked by ceiling effects.

These findings clearly have implications for our initial hypothesis that the difficulty experienced by individuals with WS on construction tasks might be accounted for by a poor or unusual ability to encode spatial relations. Construction performance represents a relative trough in ability within the visuo-spatial domain in WS. Indeed, performance on the Block Design task is significantly below that of performance on the RCPM, our current matching measure (Farran, Jarrold & Gathercole, 1999). The present results demonstrate that spatial coding is no poorer than performance on the RCPM, and therefore appear to count against our initial hypothesis.

However, our examination of the *pattern* of performance of the participant groups highlighted response biases on both of the spatial tasks that, while similar among all three TD groups, were atypical in the WS group. The consistency across typically developing groups is important to note. First, this indicates that the developmental trajectory for coding spatial relations remains stable with development; a bias evident in early childhood remains through to adulthood. Second, the differential bias observed in the WS group, is not evidence of delay as this pattern is not observed along the typical developmental trajectory, either at the developmental level of the WS group (as observed in the young TD group and matched TD group) or at the chronological age of the WS group (as observed in the adult group). These

results therefore suggest that the encoding of categorical and coordinate spatial relations is deviant in WS. This contrasts to the visual colour relations task, where no differentiation in the pattern of performance between the WS group and the matched TD controls is observed, which suggests that performance on this task is delayed rather than deviant in WS.

Even if the atypical bias observed on these spatial tasks among individuals with WS tasks represents an alternative coding strategy, it is clear that this strategy does not produce impaired levels of task performance. However, it is possible that this alternative coding strategy is less optimal than those employed by the typical population when spatial relations have to be reproduced, as in a construction task. Thus, it remains possible that poor levels of construction ability in WS are a consequence of a deviant approach to the processing of spatial relations in this group. Clearly, the next step towards demonstrating this is to determine the precise strategies employed in WS, and also to directly examine how these biases feed forward to the reproduction of spatial locations. Possible completion strategies are discussed below, after the demands of each task have been considered.

The present experiment employed spatial linguistic terms to label categories. Each category had six exemplars. Thus, participants were required to categorise each spatial location of the ball linguistically. In order to do this, the individual needed to create a non-linguistic representation of category boundaries, and category prototypes. The results, therefore, give direct insight into the linguistic coding of spatial location, and an indication of non-linguistic spatial location processing in WS. This non-linguistic coding can be related to Huttenlocher, Hedges, and Duncan's (1991) Category Adjustment Model (CAM), which emphasises the use of category boundaries and prototypes when estimating location.

In the categorical task, the individual is given the category boundary (the vertical centre of the bat) and is required to extend this horizontally in order to classify each ball position into a category. Two categories must be formed, one above the boundary and one below. According to Huttenlocher et al's model, individuals may be uncertain as to the exact category boundary, and would be more likely to correctly classify stimuli that are further away from this boundary. This was observed in the WS and matched TD groups, who became linearly more accurate and showed quicker RTs as the position of the ball was further away from the boundary (as there is no outer category boundary, the notion of increased accuracy in central category positions is irrelevant to this task). However, this pattern was not observed in the adult group or the young TD group due ceiling effects in the adult group, and the amount of variance within the young TD group.

In the co-ordinate relations task, the important information that the individual must remember is the more fine-grain information of the co-ordinates of individual balls as representing 'in' and 'out'. The dividing lines between 'in' and 'out', which were present in the practise trials only, form the category boundaries between an 'in' response and an 'out' response. Thus although categorical information is still required, success is dependent on accurate co-ordinate encoding. The performance of all four groups on this task showed linear increases in accuracy as item position moved further away from the relevant category boundaries, as predicted by the CAM.

The linear pattern of performance in the WS group indicates that they understood the demands of the tasks. A lack of understanding would be evident by a pattern of accuracy/ RT which did not relate to difficulty level, rather than the clear relationship between difficulty level and accuracy/ RT observed. However, it is clear that the systematic biases exhibited by the WS group in performance on the spatial

tasks were in the opposite direction to the remaining three participant groups. In the co-ordinate task, the WS group seemed to have underestimated the required distance from the bat to the 'in' out' division co-ordinates, thus showing the most errors on the 'in' response nearest the division points. Conversely, the matched TD, adult and young TD groups appeared to have overestimated this distance, shown by their pattern of errors, with the most errors made on the nearest 'out' position to the division points. Similarly, group differences were seen in the categorical task in the nature of response bias. The WS group appeared to have moved the category boundary (above versus below) down, with most errors made in the first position below the centre of the bat. This contrasts to the responses of the matched TD, adult and young TD groups who seemed to have moved the category boundary up, showing the most errors in the first position above the centre of the bat.

The biases observed in the two spatial tasks in this study suggest that there was uncertainty in the location of the category boundaries. In the co-ordinate task, individuals were given the categories, 'in' and 'out' by which to classify the locations of particulars. In order to remember the coordinates of the 'in'/ 'out' dividing line, participants could have used the additional category boundary between above and below which was clearly marked by the bat. It appears that the matched TD, adult, and young TD groups did indeed use this category boundary, which divided the 'in' category into two smaller, adjacent 'in' categories. Due to their uncertainty of the coordinates of the 'in'/ 'out' division line, it appears that their responses relied quite heavily on the more concrete category boundary of the bat. It is known that both typically developing children and adults overestimate distances between categories (Plumert & Hund, 2001), therefore by dividing the 'in' category into two, more particulars were classified as near to the bat, i.e. 'in' than far from the bat, 'out'.

In contrast, the individuals with WS appear to have kept to the two categories explained to them, 'in' and 'out'. As such, the position of the bat resembles a prototypical response for the 'in' category. It has been shown that typical children and adults underestimate distances between objects in the same category, typically displacing objects towards the category prototype (Plumert & Hund, 2001).

Furthermore, the CAM (Huttenlocher et al., 1991) explains that the less certain one is of the particular value, the more one's estimated particular value is weighted by the prototype. Thus, it appears that, due to uncertainty of the coordinates of the 'in'/'out' category boundary, the individuals with WS weighted their answer heavily towards the category prototype. Thus only those particulars which were very close to the bat were classified as 'in' and the remaining particulars were classified as 'out' (note that the 'out' category was disadvantaged as it had no prototypic value). Thus, the WS group were showing a contraction bias towards the category prototype (see Poulton, 1979).

The notion that individuals with WS did not divide the categories further appears reminiscent of Sandberg's (1999) study in which she measured the development of coding spatial location. She demonstrated that young children impose fewer categories on a visual array. One could argue therefore, that individuals with WS are less developed in coding spatial location than the remaining groups, thereby explaining why they only used the categories given to them. However, even the young TD group, who performed at or below the level of the WS group, appeared to spontaneously sub-divide the visual array in the same way as the adult participants. Clearly then, individuals with WS are employing a deviant method of coding coordinate spatial relations.

Explanations of the different response biases in the categorical task are less clear. It seems that the above/ below category boundary is best estimated by extending the vertical midpoint of the bat horizontally to the right. This requires precision, thus one would predict this action to be quite inexact. These categories have no outer boundaries, thus groups may have imposed outer category boundaries by using the edge of the screen. The WS group might have measured the category boundary from the bottom of the screen towards the centre, contracting their estimate, thus moving the category boundary down. Equally, the matched TD, adult and young TD groups may have measured category boundary from the top of the screen towards the centre, exhibiting a contraction bias in their estimation resulting in the boundary being moved up. However, this still cannot explain why the WS group prefer to use different reference points or landmarks than the remaining participant groups.

In reference to the colour relations task, the overall performance seems to follow a similar pattern to the spatial tasks; RT decreased and accuracy increased linearly as the stimulus hue became closer to a pure blue or a pure green hue. However, the patterns of performance between the groups do not differ which suggests that each group classified particulars into categories using the same techniques.

In summary, individuals with WS are able to encode both categorical and coordinate spatial relations at a visuo-perceptual level, albeit at a level commensurate with their generally poor visuo-spatial skills. Furthermore, the relative level of ability across the spatial tasks, and a visual relations task was comparable. One can infer from this that the poor level of ability is not specific to encoding spatial relations, but to relational encoding in general. Poor construction abilities, therefore, do not appear to be a reflection of a low level of spatial coding. However, as indicated by the unique bias in the responses of the WS group, it appears that individuals with WS do not code spatial location in a typical way; the patterns of performance demonstrate that the encoding of spatial location in WS is somewhat deviant. We propose that this deviance could be due to differential strategy use in WS, which in turn plays a part in the deviant local bias observed in the construction abilities of individuals with WS. Overall the present study offers some explanation for the uneven profile of visuo-spatial abilities in WS. Further investigation of the precise location coding strategies employed in WS, and how this affects the reproduction of these locations, will elucidate this hypothesis further.

References

Akshoomoff, N., & Stiles, J. (1996). The influence of pattern type of children's block design performance. *Journal of the International Neuropsychological Society*, 2, 392-402.

Arnold, R., Yule, W., & Martin, N. (1985). The Psychological Characteristics of Infantile Hypercalcaemia: A Preliminary Investigation. *Developmental Medicine* and Child Neurology, 27, 49-59.

Ballard, D. H., Hayhoe, M. M., Pook, P. K., & Rao, R. P. N. (1997). Diectic codes for the embodiment of cognition. *Behavioural and Brain Sciences*, 20, 723-767.

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

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

Bowerman, M. (1996). Learning how to structure space for language: A crosslinguistic perspective. In P. Bloom & M. A. Peterson & L. Nadel & M. F. Garrett (Eds.), *Language and space* (pp. 385-436). Cambridge, MA: MIT Press.

Brown, R. (1973). *A first language: The early stages*. Cambridge, MA: Harvard University Press.

Clark, H. H. (1973). Space, time, semantics, and the child. In T. E. Moore (Ed.), *Cognitive development and the acquisition of language*. New York: Academic.

Crawford, L. E., Regier, T., & Huttenlocher, J. (2000). Linguistic and non-linguistic spatial categorization. *Cognition*, 75, 209-235.

Farran, E. K., Jarrold, C., & Gathercole, S. E. (1999, September). Visuospatial cognition in Williams syndrome: An uneven profile of abilities. Paper presented at the *British Psychology Society*, Nottingham, U.K.

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*, *41*, 676-687

Hayward, W. G., & Tarr, M. J. (1995). Spatial language and spatial representation. *Cognition*, *55*, 38-84.

Hoffman, J. E., Landau, B., & Pagani, B. (2003). Spatial breakdown in spatial construction: Evidence from eye fixations in children with Williams syndrome.

Cognitive Psychology, 46, 260-301.

Huttenlocher, J., Hedges, L. V., & Duncan, S. (1991). Categories and Particulars: Prototype effects in estimating spatial location. *Psychological Review*, 98, 352-376.

Koenig, O., Reiss, L. P., & Kosslyn, S. M. (1990). The development of spatial relation representations: Evidence from studies of cerebral lateralization. *Journal of Experimental Child Psychology*, *50*, 119-130.

Kosslyn, S. M., & Koenig, O. (1992). Wet mind: The new cognitive neuroscience. Cambridge, MA: MIT Press.

Kosslyn, S. M., Koenig, O., Barret, A., Cave, C. B., Tang, J., & Gabireli, J. D. E. (1989). Evidence fro two types of spatial representations: Hemispheric

specialisation for categorical and coordinate relations. *Journal of Experimental Psychology: Human Perception and Performance*, 15, 723-735.

Kramer, J. H., Kaplan, E., Share, L., & Huckeba, W. (1999). Configural errors on the WISC-III block design. *Journal of the International Neuropsychological Society*, *5*, 518-524.

Lagers-van Haselen, G. C., van der Steen, J., & Frens, M. A. (2000). Copying strategies for patterns by children and adults. *Perceptual and motor skills*, *91*, 603-615.

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

Mervis. (1999). The Williams Syndrome Cognitive Profile: Strengths,
Weaknesses, and Interrelations among Auditory Short Term Memory, Language and
Visuospatial Constructive Cognition. In R. Fivush, W. Hirst, & E. Winigrad (Eds.),

Essays in honor of Ulric Neisser. Mahwah, NJ: Erlbaum.

Mervis, C. B., Morris, C. A., Bertrand, J., & Robinson, B. F. (1999). Williams Syndrome: Findings from an integrated Program of Research. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders: Contributions to a new framework from the cognitive neurosciences*. Cambridge, MA: MIT Press.

Morris, C. A., & Mervis, C. B. (1999). Williams Syndrome. In S. Goldstein & C. R. Reynolds (Eds.), *Handbook of Neurodevelopmental and Genetic Disorders in Children* (pp. 555-590). New York London: The Guilford Press.

Navon, D. (1977). Forest before trees: The precedence of global features in visual perception. *Cognitive Psychology*, *9*, 353-383.

Nickerson, E., Greenberg, F., Keating, M. T., McCaskill, C., & Shaffer, L. G. (1995). Deletions of the elastin gene at 7q11.23 occur in ~ 90% of patients with Williams syndrome. *American Journal of Human Genetics*, *56*, 1156-1561.

Philips, C., Jarrold, C., Baddeley, A., Grant, J., & Karmiloff-Smith, A. (2004). Comprehension of spatial language terms in Williams syndrome: Evidence for an interaction between domains of strength and weakness. *Cortex*, 40, 85-101.

Plumert, J. M., & Hund, A. M. (2001). The development of memory for location: What role do spatial prototypes play? *Child Development*, 72(2), 370-384.

Poulton, E. C. (1979). Models for biases in judging sensory magnitude. *Psychological Bulletin*, 86, 777-803.

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

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, Behavioural and Clinical Perspectives* (pp. 367-392). New York: Cambridge University Press.

Sandberg, E. H. (1999). Cognitive constraints on the development of hierarchical spatial organization skills. *Cognitive Development*, *14*, 597-619.

Shah, A., & Frith, U. (1993). Why do autistic individuals show superior performance on the block design task? *Journal of Child Psychology and Psychiatry*, *34*, 1351-1364.

Wechsler. (1981). Wechsler Adult Intelligence Scale-Revised. San Antonio, TX: The Psychological Corporation.

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

Participant group	CA(years; months): mean(SD)	RCPM: mean (SD)
WS (N=21)	21;2 (7;10)	18.00(5.13)
Matched TD (N=21)	6;3 (0;6)	17.57(5.00)
Young TD (N=20)	4;5 (0;8)	11.58 (2.24)
Adults (N=21)	21;0 (3;10)	NA

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Figure captions

Figure 1a: Categorical relations stimulus, showing each of the 12 possible ball positions.

Figure 1b: Coordinate relations stimulus, showing each of the 12 possible ball positions.

Figure 2a: Correct Responses on the Categorical spatial relations task

Figure 2b: Re-coded Correct Responses on the Categorical spatial relations task

Figure 3a: Correct Responses on the Co-ordinate spatial relations task

Figure 3b: Re-coded Correct Responses on the Co-ordinate spatial relations task

Figure 4a: Correct Responses on the Visual relations task

Figure 4b: X, Y-chromaticity co-ordinates for the colours of the stimuli employed in the visual relations task















