

RUNNING HEAD: Orientation coding and Williams syndrome

Orientation coding: A specific deficit in Williams syndrome?

Emily K. Farran

School of Psychology

University of Reading

Address correspondence to:

Emily Farran
School 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

Orientation coding: A specific deficit in Williams syndrome?

Abstract

Williams syndrome (WS) is a rare genetic disorder with a unique cognitive profile in which verbal abilities are markedly stronger than visuo-spatial abilities. The present study investigates the claim that orientation coding is a specific deficit within the visuo-spatial domain in WS. Experiment 1 employed a simplified version of the Benton Judgement of Line Orientation task, and a control, length matching task. Results demonstrated comparable levels of orientation matching performance in the WS group and a group of typically developing controls matched by non-verbal ability, although it is possible that floor effects masked group differences. A group difference was observed in the length matching task due to stronger performance from the control group. Experiment 2 employed an orientation discrimination task and a length discrimination task. Contrary to previous reports, the results showed that individuals with WS were able to code by orientation to a comparable level as their matched controls. This demonstrates that, although some impairment is apparent, orientation coding does not represent a specific deficit in WS. Comparison between Experiments 1 and 2 suggest that orientation coding is vulnerable to task complexity. However, once again, this vulnerability does not appear to be specific to the WS population, as it is also apparent in the TD controls.

Introduction

Williams syndrome (WS) is a genetic disorder which occurs in approximately 1 in 20,000 live births (Morris & Mervis, 1999). This population shows a characteristically atypical cognitive profile in which visuo-spatial abilities are markedly inferior to verbal abilities (Udwin & Yule, 1991). Furthermore, visuo-spatial performance differs substantially across areas of ability (Farran & Jarrold, 2003).

The unusual pattern of visuo-spatial abilities in WS was thought to be accounted for by a local processing bias (e.g. Bellugi, Sabo & Vaid, 1988). This pattern is observed on visuo-spatial construction and drawing tasks. Performance on such tasks is characterised by a lack of global cohesion, despite accuracy in reproducing the local elements of the visual array. The local processing bias hypothesis, however, does not hold at the level of perception. On perceptual tasks individuals with WS are sensitive to both the local and global aspects of a visual array (Farran, Jarrold, & Gathercole, 2003). Precisely why a local bias is observed in construction, but not perception, is still under investigation. Hypotheses put forward to explain this deficit include a difficulty in switching attention (Pani, Mervis, & Robinson, 1999), reduced monitoring of construction solutions (Hoffman, Landau, & Pagani, 2003), impaired encoding of spatial relations (Farran & Jarrold, submitted), and difficulty using mental imagery and coding orientation (Farran, Jarrold & Gathercole, 2001). It is possible that a clearer understanding of the relative deficits in WS, such as visuo-spatial construction and drawing, might determine the underlying reasons for the unique visuo-spatial profile in WS.

Another area in which performance is thought to be particularly poor in WS is orientation coding. This refers to any processing requirement that involves

determining the orientation of part of the visual scene, either independently or in relation to another part of the visual scene. Dupont et al. (1998), using Positron emission tomography, determined that simultaneous and successive orientation discrimination in adulthood, is a property of the right middle fusiform gyrus, the right lingual gyrus and the left middle occipital region. In addition, Slater and colleagues (Slater, Morison and Somers, 1988; Slater, Mattock, Brown & Bremner, 1991) demonstrated that the ability to code orientation is present from birth. First, newborns are able to discriminate between square wave gratings oriented from upright by 45° clockwise and gratings oriented by 45° anticlockwise (Slater et al., 1988). Second, newborns can process angular relations when presented with acute and obtuse angles (Slater et al., 1991). Thus, it appears that orientation coding abilities are available from birth. However, one cannot assume that the same brain regions are being activated in infancy as those observed in adulthood.

To my knowledge, there has been little research into the development of orientation coding beyond infancy. However, a number of anomalies have been reported: poorer performance is observed on tasks that involve oblique orientations than those that involve vertical and horizontal orientations: the 'oblique effect' (Appelle, 1972). Essock (1980) describes two classes of oblique effect. A class 1 oblique effect, present in both children and adults, refers to poorer performance due to reduced acuity or sensitivity to oblique orientations at a neural level. A class two oblique effect describes the confusion over mirror-imaged or symmetrical oblique orientations observed in young children (e.g. Rudel & Teuber, 1963). Children aged 5 to 6 years are also likely to draw oblique angles as more perpendicular to an adjoining line than they actually are (the 'perpendicular error': Ibbotson & Bryant, 1976). This is not thought to be perceptual, as the child is aware of their error. It appears to reflect

the learning of right-angled markers, which bias drawing ability until other object features are learnt. Such anomalies need to be taken into account when interpreting the WS pattern of orientation coding performance.

Evidence suggests that poor performance in WS on a number of visuo-spatial tasks could be accounted for by impaired orientation coding. For example, performance on a mental rotation task, a mental image transformation task that involves orientation coding, is significantly below the general level of visuo-spatial cognition of individuals with WS (Farran, Jarrold & Gathercole, 2001). This contrasts to performance on a mental size transformation task, an image transformation tasks which does not involve orientation coding, which is at a level commensurate with the general level of visuo-spatial ability in WS (Farran & Jarrold, 2004). One could argue that the difference in level of performance between these two tasks reflects poor orientation coding in WS.

Further evidence from a factor-by-factor analysis of the Block Design task demonstrates deviant orientation processing in WS. Participants were presented with either four squares that were divided into two halves by a diagonal line (oblique trials) or four squares that were divided in two by a horizontal / vertical line (nonoblique trials). Participants were asked to recreate a pattern in a 2 by 2 formation, using the set of four squares given to them. Whilst the controls demonstrated a class 2 oblique effect as expected, the individuals with WS did not: nonoblique and oblique trials were equally difficult, which indicates deviant orientation processing in WS (Farran & Jarrold, 2004).

Direct analysis of orientation coding in WS has been carried out using the Benton Judgement of Line Orientation Test (JLOT; Benton et al., 1978; see Bellugi, Sabo & Vaid, 1988; Rossen, Klima, Bellugi, Bihrlé, & Jones, 1996; Wang, Doherty,

Rourke, & Bellugi, 1995). A display of 11 lines oriented 18 degrees apart and two target lines of the same orientation are presented to the participant. Participants are asked to decide which of the 11 lines matches the orientation of two target lines. Bellugi et al. (1988) assessed the performance of three older children with WS. One individual did not even pass the pretest, which requires passing 2 out of 5 practice trials. The remaining two WS participants scored at or below 35% correct. Wang et al. (1995) assessed 10 individuals with WS (mean age: 15.7 years). Eight of this group also failed the pretest. Rossen et al. (1996) assessed 6 individuals with WS using this task (mean age 14;2) with similar results; the majority of the group did not pass the pre-test. These results demonstrate striking floor effects, i.e. the majority of individuals were unable to complete even the lowest level of the task. Clearly, this is not an appropriate task by which to measure orientation coding in WS; it does not tell us the actual level of orientation coding ability in WS, but informs us that they are at some point below the lowest level measured by this task.

Stiers, Willekens, Borghgraef, Fryns, & Vandenbussche (conference proceedings, 2000) designed a version of the JLOT which displayed a reduced number of choice alternatives as a way of simplifying the task: the Pre-school Judgement of Line Orientation task (PJLO). Individuals were asked to identify a target amongst distracters. In blocks 1 to 3, one target line was presented and the number of choice alternatives ranged from 2 alternatives in block 1, to 4 in block 2, and 11 alternatives in block 3. Block 4 used items from the original Benton lines task, with 2 target lines and 11 response choices. Twenty individuals with WS of ages ranging from 5 to 25 years performed at a level that was slightly below their verbal ability, and at the same level as their non-verbal ability as measured by the Wechsler Pre-school and Primary Scale of Intelligence- Revised (WPPSI-R; Wechsler, 1989).

Although no level of orientation matching ability was given, the results suggest that individuals with WS are able to encode differences in line orientations when the number of choice alternatives is reduced.

Experiment 1

Experiment 1 is an attempt to identify the level of orientation coding ability in WS using a design similar to the PJLO (Stiers et al., 2000) and the JLOT (Benton et al., 1978). Performance will be measured relative to typically developing (TD) controls of the same general level of visuo-spatial ability, but also relative to a control task. The control task, a length matching task, has the same task demands as the orientation task, but without the orientation coding factor. This enables one to determine the extent to which the orientation demands affect performance over and above any other task demands, such as making a choice from distracters or segmenting stimuli from the general pattern of the display.

Method

Participants

Thirteen individuals with WS were recruited from the records of the Williams syndrome foundation, UK. All individuals had been positively diagnosed with WS using phenotypic and genetic information. Genetic diagnosis was by a Fluorescent insitu Hybridisation (FISH) test. This checks for the deletion of elastin on the long arm of chromosome 7, which occurs in approximately 95% of individuals with WS (Lenhoff, Wang, Greenberg & Bellugi, 1997). The WS group were matched individually by visuo-spatial ability (non-verbal reasoning), to 13 typically developing (TD) children using 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. Table 1 illustrates the RCPM raw scores, and chronological age of each group.

Table 1 about here

Design and Procedure

Two tasks measured orientation and length matching respectively, using a threshold procedure. Each task comprised of 4 trials at each of 5 levels. These levels had 2, 4, 6, 8, and 10 choice alternatives. Both tasks were administered to participants in one testing session along with two other visuo-spatial tasks not presented here, with order of presentation counterbalanced.

Participants were shown images on an A4 page of a booklet. Participants were asked to point to the choice stimulus that they thought matched the target on the dimension (orientation or length) being assessed. There was no time limit. The experimenter noted their response, and recorded accuracy. Participants proceeded to the next level if they gave at least 3 out of 4 correct responses. The dependent variable was the participant's threshold level of ability (0 to 5), determined as the final level at which the above criteria were met.

Figures 1 and 2 about here

Orientation task.

The orientation task used a similar procedure to the JLOT. Each A4 page displayed a target line on the left and a set of choice stimuli on the right. Choice stimuli were spaced to cover an angle which was less than 90°. This was to eliminate any potential problems that might occur if including lines which were symmetrical about a horizontal or vertical axis such as class 2 oblique effects (Essock, 1980, see Farran & Jarrold, 2004). Lines were displayed in a fan formation subtending an

overall angle of 30, 55, 60, 65, and 70 degrees for levels 1 to 5 respectively. As such, the orientation between choice stimuli was; 30° (level 1, 2 lines), 18.33° (level 2, 4 lines), 13° (level 3, 6 lines), 10° (level 4, 8 lines), and 8.33° (level 5, 10 lines) (Figure 1).

Length task.

The length task was designed to mirror the orientation task where possible. Thus, each A4 page displayed a target line on the top left of the page and a set of choice stimuli diagonally below, to the right. Horizontal lines were displayed one above the other, half of the trials in ascending and half in descending order of length. Lengths of lines were determined in pixels, but are described as they appeared on the A4 page, in mm. Lines differed in length by 67mm (2 lines), 23mm (4 lines), 13mm (6 lines), 9mm (8 lines), and 7mm (10 lines) for levels 1 to 5 respectively. In order to cover a uniform area of space at each level, vertical spacing between choice stimuli also varied from 89mm (level 1) to 11mm (level 5) (Figure 2).

Results

Performance was poor in the orientation task, with floor effects from four individuals with WS and 4 control children (these children did not pass level 1). Rather than removing these participants and their matched pair, floor performance was given a threshold of 0. No floor effects were observed in the length task.

Nonparametric analyses were carried out. This was because one could argue that the dependent variable is not strictly continuous, and because the WS length data and the TD orientation data were not normally distributed (WS length: Kolmogorov-Smirnov $z = 1.69$, $p = .01$; TD orientation: Kolmogorov-Smirnov $z = 1.25$, $p = .09$). Mann-Whitney U and Wilcoxon tests were employed to analyse the effect group (WS, TD) and task (orientation, length) respectively. This revealed no significant

group differences: orientation performance, $U=71$, $N_A=13$, $N_B=13$, $p=.45$; length performance, $U=57$, $N_A=13$, $N_B=13$, $p=.10$. There was a significant effect of task for the TD group ($T=4.5$, $N=13$, $p = .01$), due to superior performance on the length, compared to the orientation task. The effect of task was not significant for the WS group ($T=15.0$, $N=13$, $p = .86$). Results are illustrated in Figure 3.

Figure 3 about here

Discussion

Previous research demonstrated that individuals with WS typically perform at floor on the JLOT (e.g. Bellugi et al., 1988). Stiers et al. (2000) demonstrated using a simpler version of the task, that WS performance was at a level expected of their non-verbal mental age measured by the WPSSI. The present results also show no difference between the WS group and visuo-spatial matched TD controls on an orientation task. However, floor performance was observed in 4 members of each group, WS and TD, and many individuals passed level 1 (2 choice alternatives) only (WS: $N=5$; TD: $N=8$). Thus, the task may be failing to differentiate between WS and control group performance. Sufficient details are not given to determine if a similar effect occurred in the study by Stiers et al. (2000).

The length task shares all task demands, but the orientation coding factor, with the orientation task. Due to the relatively elevated performance on the length task in the TD controls, one can be confident that the poorer performance on the orientation task in this group relates to the orientation coding requirement, and thus, for typically developing children, matching by shared orientation is harder than matching by the factor of length. The flat profile of the WS group is difficult to interpret; performance on the two tasks could be at a similar level due to their shared factors, or due to the

separable factors of orientation coding and length coding respectively (which would imply a relative deficit in length coding in WS). However, since the floor effects in the orientation coding task may be masking poorer abilities on this task, which would indicate an overall delay in WS, the flat WS profile is not explored further.

Orientation coding and length coding are examined using an alternative methodology in Experiment 2.

Experiment 2

Where Experiment 1 predominantly focused on the number of choice alternatives as an index of difficulty, Experiment 2 employs a discrimination design, which focuses more specifically on the increment of difference (degrees of orientation / mm of length) between stimuli. This enables one to determine the point at which the difference between stimuli becomes detectable to the individual, thus reducing the likelihood of floor effects. Two tasks investigate the ability to code by orientation and by length respectively; the individual is shown two lines and asked if they are the same or different.

Method

Participants

Experiment 2 took place approximately 20 months after Experiment 1. Seventeen individuals with WS were employed, 11 of whom had participated in Experiment 1, and a further six who were recruited from the records of the Williams Syndrome Foundation, UK. All individuals had been positively diagnosed with WS using phenotypic and genetic information. Genetic diagnosis was by a Fluorescent insitu Hybridisation (FISH) test. As in Experiment 1, the WS group were matched by visuo-spatial ability (non-verbal reasoning) to TD children using the Ravens Coloured

Progressive Matrices (RCPM; Raven, 1993). Table 2 illustrates the RCPM raw scores, and chronological ages of each group.

Table 2 about here

Design and Procedure

Tasks were presented on a laptop computer. There were two response pads, a large green tick for 'same' responses and a large red cross for 'different' responses. Stimuli remained on the screen until the correct response had been given, at which point a mask appeared for 300msecs, followed by the next trial.

Figures 4 and 5 about here

Orientation discrimination task.

Participants were told that two lines would appear on the screen, one on the left and one on the right, whose orientation would either be the same or different (Figure 4). This was presented to participants in a way that the participants could understand by describing the lines as arrows, or by using hand movements to indicate orientation. It was explained that they were to press the green tick if they thought that the lines were the 'same' and the red cross for a 'different' response. There were four practice trials, 2 same trials and 2 different trials (the easiest two levels: lines differed by 55° and 60°). Experimental trials began once the experimenter was confident that the participant understood the procedure. In practise, no participants required more than these 4 practice trials.

There were 96 experimental trials; 48 same and 48 different trials. Four versions of same trials were employed; two lines, left and right, both oriented 10° or 20° anticlockwise from horizontal, and the mirror reverse of these two trial types. The different trials were created using the non-reversed same trials; the left line remained

as in the same trial (10° or 20°), whilst the right line was oriented a further 5° to 60° . These trial types were also mirror reversed to create a further two trial types. There were 12 increments of difference in orientation ranging from 5° to 60° at 5-degree intervals. Thus, in accord with the same trials, there were four types of each of the 12 increments of difference in orientation (4 x 12 trials).

Experimental trials were run in 4 blocks of 24 trials. Each block consisted of 12 same and 12 different trials (one at each increment of difference), each of which comprised 6 original and 6 mirror reversed trials. Increments of orientation difference alternated between original and mirrored trials. Trials were randomised within each block.

Length discrimination task.

Participants were shown two horizontal lines and asked if they were the same or different in length (Figure 5). As above, they were asked to press the tick or cross for same and different responses respectively. As above, all participants demonstrated task comprehension during the four practice trials (two same, two different).

All lengths were calculated in pixels. On the laptop employed, $1\text{mm} = 3.28$ pixels. In same trials, there were two lines, 100 pixels (30.5mm) in length with a 50 (15.3mm) or 200 (61mm) pixel gap between their near ends. Different trials were created from these trials by extending the left or right line away from (50 pixel gap trials) or towards (200 pixel gap trials) the centre. This created 4 versions of each difference in length. Differences ranged from 10 to 100 pixels at 10 pixel (3.1mm) increments. The difference between the two lines of X pixels is henceforth known as an 'Xdiff' trial.

Experimental trials were run in 4 blocks of 20. Each block consisted of 10 same and 10 different trials of the same trial type (either 50 or 200 pixel gap trials),

one trial at each increment of difference. Left or right line extension alternated with increments of differences in length. Trials within each block were randomised.

Results

Data were analysed in terms of proportion of 'different' responses and response times (RT) to correct responses for both tasks separately. Participants were removed if their False Positive (FP) responses were higher than their Hit Rate (HR) on 50% or more levels (6 or more levels of orientation difference, or 5 or more levels of length difference). This was apparent in the performance of one individual with WS in the orientation discrimination task, and one individual with WS in the length discrimination task. For each task analysis, the data of the WS individual and their matched control were removed.

Orientation discrimination

Response times.

For cells in which participants achieved zero out of four correct responses, a RT value was given, taken from the participant group mean at that level. This is a conservative method as it credits failed responses with a RT typical of a successful response. Any significant group differences are therefore unlikely to result from this adjustment to the data, as the adjustment would bring level of ability closer together. The distribution of some of the data points was significantly different from a normal distribution (WS, $p < .05$ for 45°, 55°, 60°; TD, $p < .05$ for 15°). Some distributions of performance were also skewed (WS: $z > 1.96$ for 0°, 5° and 15° to 60°; TD: $z > 1.96$ for 5° to 25°, 35° and 40°). Thus, a logarithmic transformation was used to ensure that parametric assumptions were met.

A two-way ANOVA was carried out with group as the between participant factor (2 levels; WS, TD) and orientation as the within participant factor (13 levels; 0°

to 60° difference). The main effect of group was not significant, $F < 1$. There was a significant main effect of orientation, reported as a linear contrast, $F(1, 30) = 12.93$, $p = .001$, partial $\eta^2 = .30$. This was due to reaction times decreasing linearly as the difference in orientation became greater. The interaction was not significant, $F < 1$.

Proportion of 'different' responses.

Figures 6 and 7 illustrate proportion of 'different' responses and d prime respectively. D prime is arguably a more sensitive measure of performance than the proportion of different responses, as it takes participant's biases into account.

Calculation of d prime requires that hit rates (HR) and false positive (FP) values are not 1 or 0. Where values were 1, the proportion correct was replaced by $1 - 1/(2N)$, and where values were 0, the proportion correct was replaced by $1/(2N)$ (see Wixted & Lee, 2005). In cases where FP values were higher than the HR values, a d prime cannot be calculated. (as noted above, if this occurred on 50% or more levels, the participant data was removed). If this occurred on harder trials, before the individual had achieved a d prime of 1.35 (the equivalent to a difference in orientation which could be discriminated with probability of 75%, see Johnson, 1980), a d prime value of 0 was given. If this occurred once a d prime value of 1.35 had been reached, the d prime value was taken from an average of the d prime value of the levels above and below the missing d prime value.

D prime data was normally distributed ($p > .05$) and symmetrical ($z < 1.96$). A two-way ANOVA was carried out on d prime values, with group as the between participant factor (2 levels: WS, TD) and orientation as the within participant factor (12 levels, 5° to 60°). The main effect of group was not significant, $F < 1$. There was a significant main effect of orientation, $F(11, 330) = 23.30$, $p < .001$, partial $\eta^2 = .44$. This was due to an initial linear increase in d-prime (5° < 10° to 60°, 10° < 15° to 60°, 15° <

20° to 60° $p < .05$ for all) followed by a flattening out of d' prime (20° to 60°, $p > .05$ for all comparisons). The interaction was not significant, $F(12, 330) = 1.03$, $p = .42$, partial $\eta^2 = .03$.

Figures 6 and 7 about here

Length discrimination

The data from one WS participant was lost due to computer error for this task only. As such the data from this participant's matched control has been removed.

Reaction times.

As with the data from the orientation discrimination task, for cells in which participants achieved zero out of four correct responses, a RT value was given, taken from the participant group mean at that level. The distribution of performance followed a normal distribution ($p > .05$ for all), but displayed some skewed distributions of performance (WS: $z > 1.96$ for 10 to 20 diff, 60diff and 80 to 100diff ;TD: $z > 1.96$ for 0 to 30 diff and 50 to 100 diff). Thus, a logarithmic transformation was used to ensure that parametric assumptions were met.

A two-way ANOVA was carried out with group as the between participant factor (2 levels; WS, TD) and length as the within participant factor (11 levels: 0 to 100 pixels of difference in length). The main effect of group was not significant, $F(1, 28) = 1.42$, $p = .24$, partial $\eta^2 = .05$. There was a significant main effect of length, reported in terms of linear contrasts, $F(1, 28) = 10.74$, $p = .003$, partial $\eta^2 = .28$. This was due to a reduction in RT as the difference in length became greater. There was also a significant group by length interaction, $F(10, 280) = 4.02$, $p < .001$, partial $\eta^2 = .13$. Further exploration revealed that the effect of length was present in the WS group

only (WS: reported as a linear contrast, $F(1, 14) = 11.72, p=.004$, partial $\eta^2=.46$; TD: $F<1$).

Proportion of 'different' responses.

Figures 8 and 9 illustrate proportion of 'different' responses and d prime respectively. For d prime data, HR and FP values of 1 and 0 were replaced using the same method as in the calculations for orientation above. Similarly, the same criteria were used for cases where the FP value was higher than the HR.

D prime data was normally distributed ($p>.05$) and symmetrical ($z<1.96$). A two way ANOVA was carried out on the d prime data, with group as the between participant factor and length as the within participant factor (10 levels: 10diff to 100diff). The main effect of group was not significant, $F<1$. The main effect of length was significant, $F(9, 252)=31.96, p<.001$, partial $\eta^2=.53$. This was due to an initial linear increase in d prime with difference in length: 10diff > 20 to 100diff, 20diff > 30 to 100 diff, 30 diff > 50 to 100diff, 40diff >90 to 100diff, 50diff > 100 diff ($p<.05$ for all). This slope flattened for the largest differences in length: 60diff to 100diff ($p>.05$ for all comparisons). The interaction was not significant, $F(9, 252)=1.33, p=.22$, partial $\eta^2=.05$.

Discussion

The results of Experiment 2 demonstrated no overall group differences in orientation coding or in length coding. The TD controls showed some differentiation from the WS group in their pattern of RT performance on the length discrimination task. That is, level of difficulty did not affect RT in the TD group, but increased linearly with increased difficulty in the WS group. One could argue from this that the TD group was less affected by the length discrimination differences than the WS

group. However, this interaction was not mirrored in the d' prime data, a more sensitive measure of performance, and so does not appear to hold much weight.

The results do not support previous studies, which suggest that orientation discrimination is a specific impairment in WS. Indeed, the only differences observed are in the pattern of RT performance on the *length* task. Overall, the results suggest a general perceptual difficulty in perceiving small differences between objects along a single dimension, which is commensurate with the overall level of visuo-spatial impairment in WS (as measured here by the RCPM).

General discussion

It is reported that the ability to code orientation is a specific deficit within the WS visuo-spatial cognitive profile (e.g. Bellugi, Sabo & Vaid, 1988; Rossen et al., 1996; Wang et al., 1995). However, studies to date have employed the JLOT, which is simply too difficult for this population. Experiment 1 followed the design of the JLOT, but with fewer choice alternatives in an effort to simplify the task. Results showed that even this task produced floor effects, not only in the WS population, but also in the control group of typically developing five-year-olds. This suggests that this method of measuring orientation coding is too complex for individuals of this level of visuo-spatial ability. The procedure involves comparing a target line to an array of lines, thus individuals must be able to segment each line from the overall pattern of stimuli, and then make the comparison between the target and each choice alternative in isolation, without being distracted by the other choice alternatives.

Perhaps the results of Experiment 1 indicate that orientation coding ability is affected by task complexity. If orientation coding is vulnerable, it may suffer more than other visuo-spatial factors as a function of how complex the task is. Due to floor effects, the orientation task did not differentiate between the performance of the WS

group and TD controls. Nevertheless, a specific vulnerability to orientation coding in WS is supported by the difficulties experienced by individuals with WS on complex tasks such as mental rotation and the Block Design task, where difficulty with the orientation aspects of the tasks are reported (Farran, Jarrold & Gathercole, 2001; Farran & Jarrold, 2004).

Experiment 2 employed a discrimination design. This type of design is relatively less complex than the task employed in Experiment 1. Contrary to expectation, overall group differences were not observed in the orientation task. This was also apparent in the d' analysis of the length discrimination task, which suggests that orientation discrimination in WS is no more impaired than other forms of discrimination, such as length discrimination. These results highlight that, at least on low-level tasks, a deficit in stimulus discrimination is not specific to discriminating by orientation in WS. Importantly, the results demonstrate that individuals with WS can code orientation, which has implications for the claims made from the results of WS performance on the JLOT. This concern is highlighted by comparing the results of Experiments 1 and 2. In Experiment 1, nine of the thirteen individuals with WS and 12 TD controls either failed to pass level 1 or passed level 1 only. At levels 1 and 2, the angular difference between the choice alternatives was 30° and 18.33° respectively. In contrast, in Experiment 2, performance at similar differences was strong: 30° difference, WS = 82%, TD = 87% accuracy, 20° difference, WS = 77%, TD = 78% accuracy. This clearly illustrates that the results of Experiment 1, and thus the results of those studies which have employed the JLOT, do not truly represent level of orientation coding ability. Worryingly, this is not only the case in the WS population where you might expect some deviation on a visuo-spatial task, but also in the TD controls.

The present results demonstrate that the ability to code orientation in WS is poor, but available to individuals with WS. It is also no poorer than the ability to code differences in length in WS. We do not know, on account of the floor effects in Experiment 1, whether orientation coding is more vulnerable than coding by length on more complex tasks. It is entirely possible that length discrimination abilities are equally vulnerable. However, whilst orientation coding is a fundamental factor in a number of visuo-spatial tasks, particularly those which involve manipulation (perceptually or manually), coding for length is rarely an important factor in task completion. This in itself could explain why orientation coding has often been recorded as a specific deficit in WS.

The prominence of orientation coding within the visuo-spatial domain highlights the importance of systematic investigation of this factor. This study goes some way to determining the extent of this deficit in WS. However, there are many other aspects of orientation coding which may be impaired to a greater or lesser extent than orientation discrimination in WS. These aspects may influence many features of visuo-spatial cognition in WS. Farran & Jarrold (2004) have demonstrated some deviance in WS when discriminating between mirror-imaged oblique lines compared to nonoblique lines. Furthermore, aspects of mental imagery which involve orientation coding appear to be impaired relative to other mental imagery tasks (Farran, Jarrold, & Gathercole, 2001; Farran & Jarrold, 2004).

In contrast to the studies above, visuo-spatial tasks which are based on object recognition, rather than production, matching or manipulation, do not appear to be affected by orientation factors. The Benton Test of Facial Recognition (Benton, Hamsher, Varney, & Spreen, 1983) includes faces presented front-view and three-quarter view. This manipulation does not appear to impair WS performance (e.g.

Karmiloff-Smith, 1997; Rossen et al., 1996). Similarly, the Canonical-noncanonical Views Test (Carey & Diamond, 1990) involves recognition of objects from different viewpoints. Individuals with WS do not seem to be adversely effected by these differences in orientation (Wang et al., 1995).

The studies above demonstrate that a poor ability to carry out more complex visuo-spatial tasks might be accounted by orientation coding factors. Clearly further investigation is required to determine which variables are affected by orientation demands. At present it appears that object and face recognition are not adversely affected, but that manipulation and construction tasks might be. In conclusion, I suggest that orientation coding is a vulnerable ability, and thus becomes more problematic as a function of task complexity. Further investigation is required to support this suggestion, and to determine whether this is a unique feature of WS, or a characteristic of individuals at this level of visuo-spatial cognition.

References

- Appelle, S. (1972). Perception and discrimination as a function of stimulus orientation: the oblique effect in man and animals. *Psychological Bulletin*, 78, 266-278.
- Bellugi, U., Sabo, H., & Vaid, J. (1988). Spatial deficits in children with Williams syndrome. In J. Stiles-Davis & U. Kritchevsky & U. Bellugi (Eds.), *Spatial Cognition: Brain Bases and Development* (pp. 273-297). Hillsdale, New Jersey: Lawrence Erlbaum.
- Benton, A. L., Varney, N. R., & Hamsher, K. d. (1978). Visuospatial judgement: A clinical test. *Archives of Neurology*, 35, 364-367.
- Benton, A. L., Hamsher, K., Varney, N. R., & Spreen, O. (1983). *Contributions to neuropsychological assessment*. New York: Oxford University Press.
- Cecala, A. J., & Garner, W. R. (1986). Internal frame of reference as a determinant of the oblique effect. *Journal of Experimental Psychology: Human Perception and Performance*, 12, 314-323.
- Carey, S., & Diamond, R. (1990). *Canonical-noncanonical views test.*: Unpublished.
- Dupont, P., Vogels, R., Rosier, A., Cornette, L., Bormans, G., Mortelmans, L., & Orban, G. A. (1998). Regions in the brain activated by simultaneous orientation discrimination: a study with positron emission tomography. *European Journal of Neuroscience*, 10, 3689-3699.
- Essock, E. A. (1980). The oblique effect of stimulus identification considered with respect to two classes of oblique. *Perception*, 9, 37-46.
- Farran, E.K., & Jarrold, C. (submitted) Evidence for Impaired Spatial Location Coding in Williams Syndrome: An Explanation for the Local Bias in Visuo-Spatial Construction Tasks? *Manuscript submitted for publication, Brain and Cognition*

- Farran, E.K., & Jarrold, C. (2004). Exploring Block construction and mental imagery: Evidence of atypical orientation discrimination in Williams syndrome. *Visual Cognition*, 8, 1019-1040.
- Farran, E.K. & Jarrold, C. (2003). Visuo-spatial cognition in Williams syndrome; Reviewing and accounting for the strengths and weaknesses in performance. *Developmental Neuropsychology*, 23, 175-202
- 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.
- 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.
- 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.
- Johnson, K. O. (1980). Sensory discrimination: decision process. *Journal of Neurophysiology*, 43, 1771-1792.
- Lenhoff, H. M., Wang, P. P., Greenberg, F., & Bellugi, U. (1997). Williams syndrome and the brain. *Scientific American*, 277(6), 42-47.
- 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.
- Pani, J. R., Mervis, C. B., & Robinson, B. F. (1999). Global Spatial Organization by Individuals with Williams Syndrome. *Psychological Science*, 10, 453-458

- Raven, J. C. (1993). *Coloured Progressive Matrices*. Oxford, UK: Information Press Ltd.
- Rossen, R., Klima, E. S., Bellugi, U., Bihrlé, 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.
- Rudel, R. G., & Teuber, H.-L. (1963). Discrimination direction of line in children. *Journal of Comparative and Physiological Psychology*, *56*, 892-898.
- Slater, A., Morison, V., & Somers, M. (1988). Orientation discrimination and cortical function in the human newborn. *Perception*, *17*, 597-602.
- Stiers, P., Willekens, D., Borghgraef, M., Fryns, J. P., & Vandenbussche, E. (2000). *Visuo-perceptual and visuo-constructive ability in persons with Williams syndrome*. Paper presented at the 8th International Professional Conference on Williams Syndrome, Dearborn, Michigan.
- Slater, A., Mattock, A., Brown, E., & Bremner, J. G. (1991). Form perception at birth: Cohen and Younger (1984) revisited. *Journal of Experimental Child Psychology*, *51*, 395-406.
- Visuo-perceptual and visuo-constructive ability in persons with Williams syndrome*. Paper presented at the 8th International Professional Conference on Williams Syndrome, Dearborn, Michigan.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, *13*, 232-244.
- 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.

Wechsler, D. (1989). *Wechsler Pre-school and Primary Scale of Intelligence-Revised*. New York: The Psychological Corporation.

Wixted, J. & Lee, K. (2005). <http://psy.ucsd.edu/~kang/sdt/sdt.htm>

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 (years; months)	RCPM score
	Mean (S.D.)	Mean (S.D.)
WS	21;8 (11;1)	17.85 (4.18)
TD	5;8 (0;6)	18.23 (3.85)

Table 2: Participant details

Group	CA (years; months)	RCPM score
	Mean (S.D.)	Mean (S.D.)
WS	21;9 (11;1)	17.59 (6.71)
TD	5;10 (0;5)	16.94 (6.76)

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. 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 Captions

Figure 1: Experiment 1: Orientation task stimuli

Figure 2: Experiment 1: Length task stimuli

Figure 3: Orientation and Length matching performance: Means (S.E.)

Figure 4: Experiment 2: Orientation task stimuli

Figure 5: Experiment 2: Length task stimuli

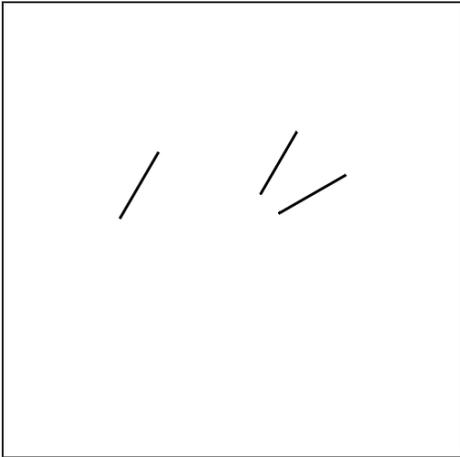
Figure 6: Orientation task, proportion of 'different' responses: Means (S.E.)

Figure 7: Orientation task, d prime values: Means (S.E.)

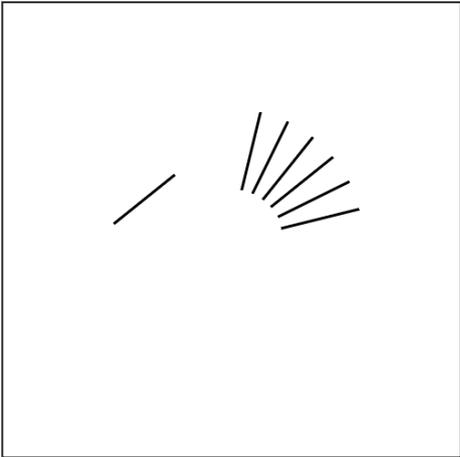
Figure 8: Length task, proportion of 'different' responses: Means (S.E.)

Figure 9: Length task, d prime values: Means (S.E.)

Figure 1

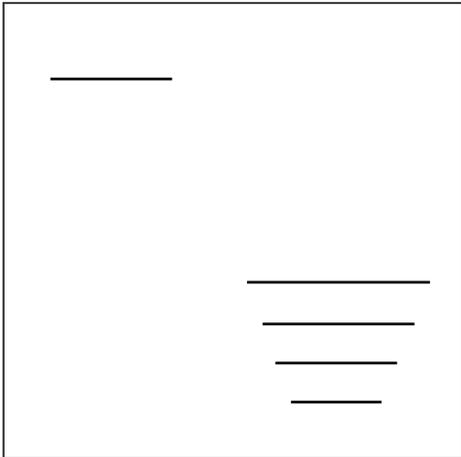


Level 1 (2 alternatives)

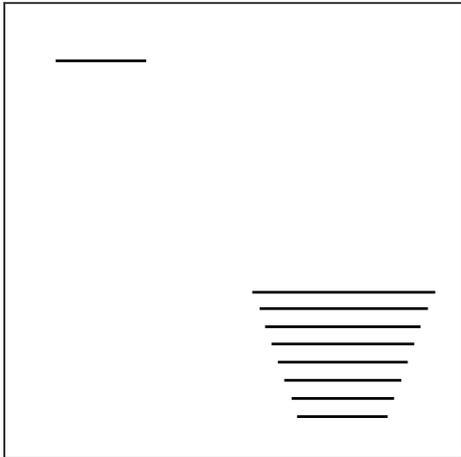


Level 3 (6 alternatives)

Figure 2



Level 2 (4 alternatives)



Level 4 (8 alternatives)

Figure 3

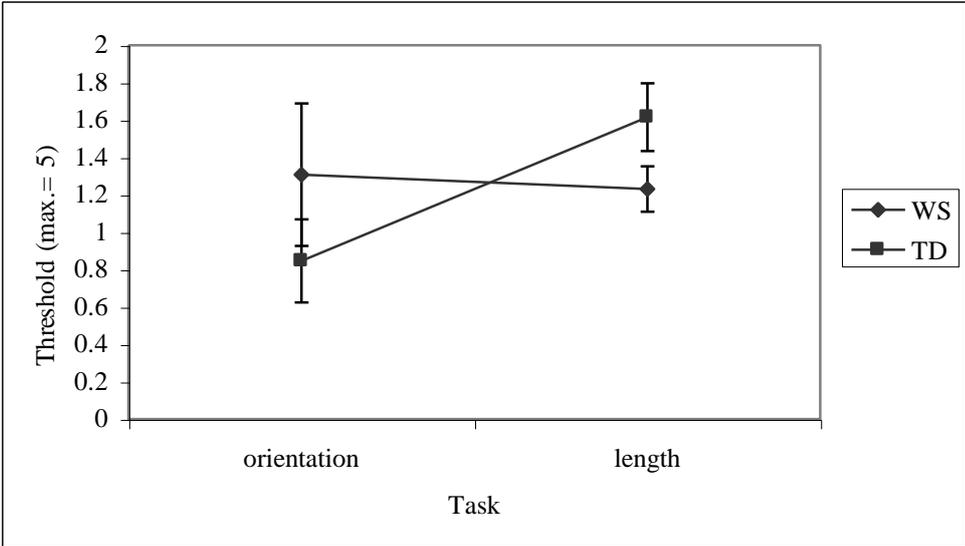
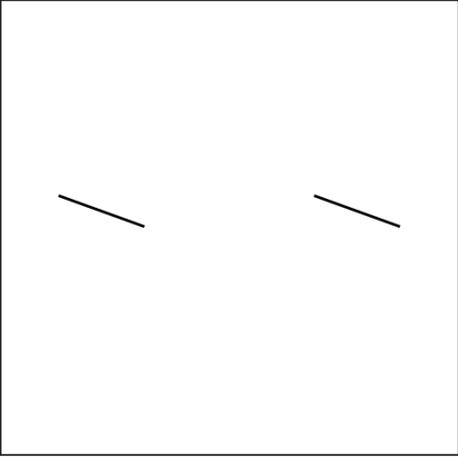
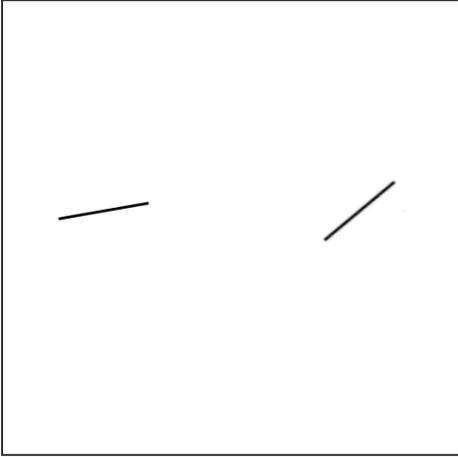


Figure 4

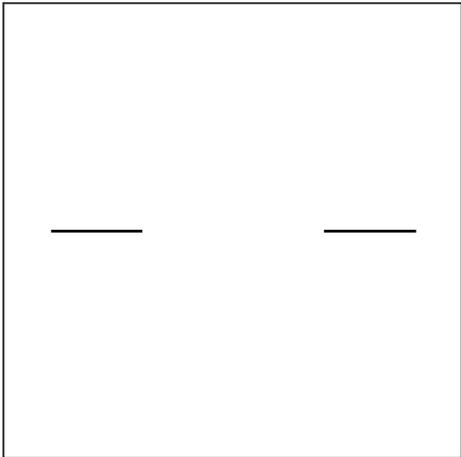


Same trial

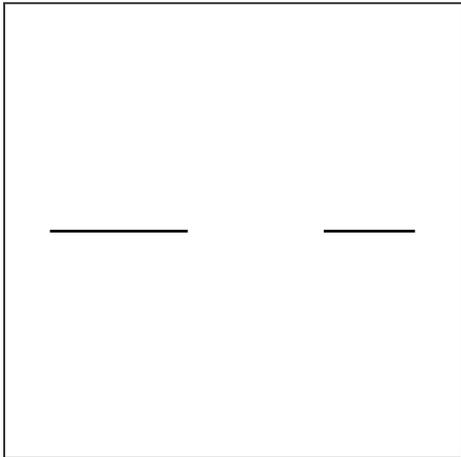


Different trial (30° difference)

Figure 5



Same trial



Different trial (40 pixel difference)

Figure 6

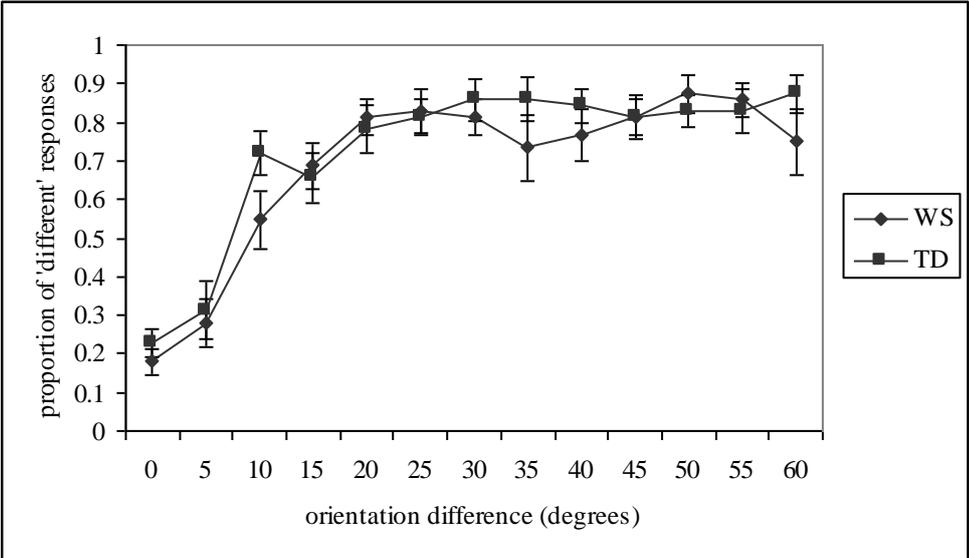


Figure 7

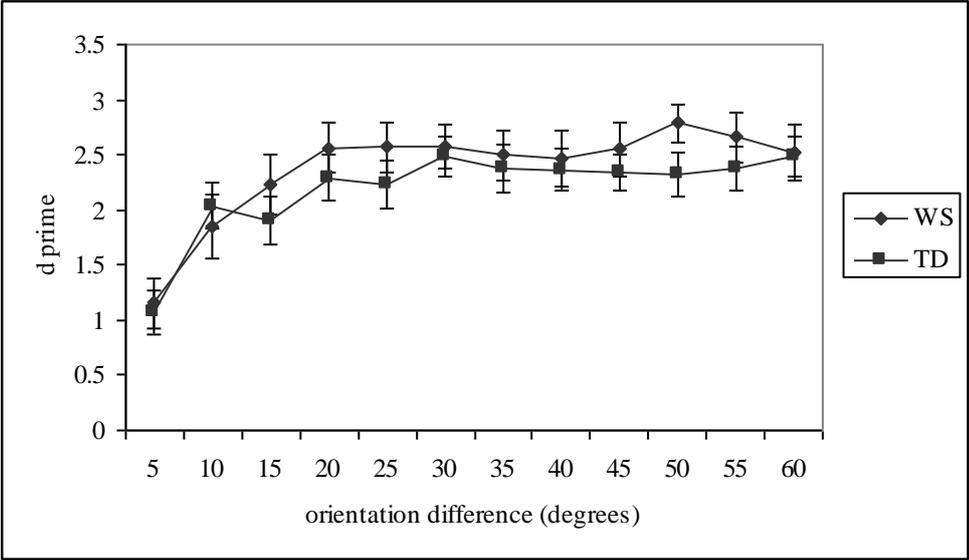


Figure 8

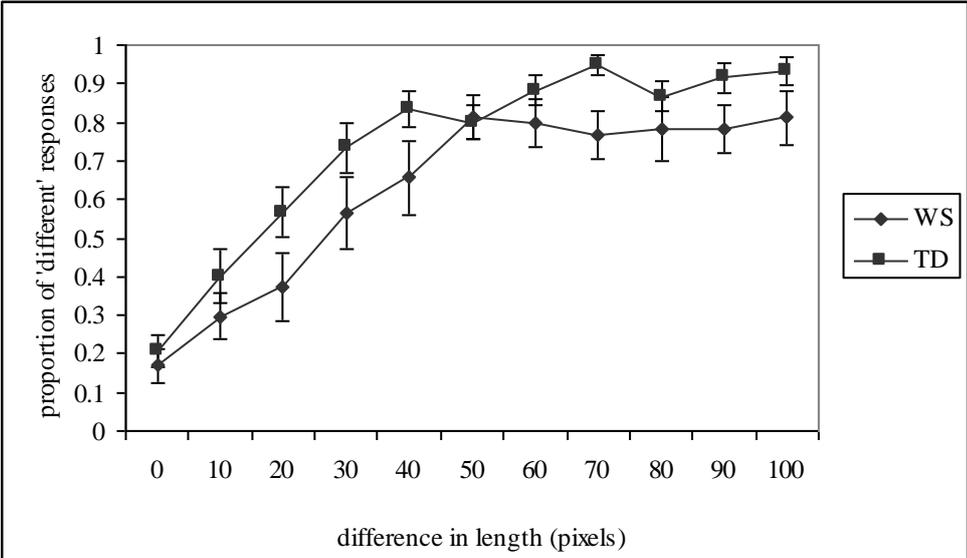


Figure 9

