Exploring Block Construction and Mental Imagery: Evidence of atypical orientation discrimination in Williams syndrome

Emily K. Farran
University of Reading

Christopher Jarrold
University of Bristol

Short Title: ORIENTATION DISCRIMINATION IN WS

Address correspondence to:

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

The visuo-spatial perceptual abilities of individuals with Williams syndrome (WS) were investigated in two experiments. Experiment 1 measured the ability of participants to discriminate between oblique and between nonoblique orientations. Individuals with WS showed a smaller effect of obliqueness in response time, when compared to controls matched for non-verbal mental age. Experiment 2 investigated the possibility that this deviant pattern of orientation discrimination accounts for the poor ability to perform mental rotation in WS (Farran et al., 2001). A size transformation task was employed, which shares the image transformation requirements of mental rotation, but not the orientation discrimination demands. Individuals with WS performed at the same level as controls. The results suggest a deviance at the perceptual level in WS, in processing orientation, which fractionates from the ability to mentally transform images.
Exploring Block Construction and Mental Imagery: Evidence of atypical orientation discrimination in Williams syndrome

**Introduction**

Williams syndrome (WS) is a rare genetic disorder occurring in approximately 1 in 25,000 births (Morris & Mervis, 1999), that is characterised by a particular cognitive phenotype in which visuo-spatial abilities are impaired in comparison to relatively superior verbal abilities (e.g. Mervis, Morris, Bertrand, & Robinson, 1999). Two dominant hypotheses have been put forward to explain the deviance in visuo-spatial cognition. The first, the local processing bias hypothesis (e.g., Bellugi, Sabo & Vaid, 1988), asserts that individuals with WS prefer to attend to the parts or details of an image, rather than the image as a whole. In fact, individuals with WS do show a local bias in drawing and construction tasks where they tend to sacrifice global accuracy for local detail, but not on perceptual tasks, where the balance of local and global processing appears to be typical in WS (Farran & Jarrold, 2003; Farran, Jarrold & Gathercole, 2001, 2003; Pani, Mervis, & Robinson, 1999).

The second hypothesis relates to differences in the function of the ventral and dorsal visual streams in WS, which are thought to be responsible for perception and action respectively (Milner & Goodale, 1995). Atkinson et al. (1997) demonstrated a relative impairment in WS performance on a ‘dorsal stream’ post box task, in which the individual posts a card into a slot, compared to a ‘ventral stream’ version of the task, where the participant must indicate the orientation of the posting slot. As a consequence they suggested that individuals with WS have a deficit in dorsal stream processing, which might explain their difficulties in drawing and construction, although they also noted that “…their pattern of deficit, make it unlikely that a dorsal
stream deficit is the only neural basis for cognitive and performance anomalies in WS.” (p. 1921). It therefore appears that there may be a number of factors that contribute to the profile of visuo-spatial abilities in WS.

These two competing hypotheses can be investigated by examining the factors that constrain performance on the Block Design task of the Wechsler scales (e.g., Wechsler, 1974, 1981). Individuals with WS typically perform less well on this task than on other visuo-spatial tasks such as the Picture Completion and Picture Arrangement subtests of the Wechsler Intelligence Scale for Children (WISC; Wechsler, 1974) and the Children’s Embedded Figures test (CEFT; Witkin, Oltman, Raskin, & Karp, 1971) (e.g., Dall’oglio & Milani; Farran et al. 2001; Udwin & Yule, 1991), despite the fact that these other tasks themselves already show a substantial impairment relative to verbal ability (e.g., Arnold, Yule & Martin, 1985; Howlin, Davies & Udwin, 1998). In the Block Design task, the participant must put a number of blocks together so that the upper faces resemble a model image. This involves two major stages; individuals must first perceptually segment the model image into component parts and second, integrate these to resemble the model image (using both visuo-motor and perceptual processes) (see Kohs, 1923). These two stages both require processing at local and global levels, hence why, through manipulation of the local processing requirements, the Block Design task is known to index extent of local processing bias (e.g., Shah & Frith, 1983). Both stages may well also involve mental imagery – a property of the dorsal visual stream (Alivisatos & Petrides, 1997; Larsen, Bundesen, Kyllingsbaek, Paulson, & Law, 2000).

Preliminary support for the suggestion that a local processing bias in WS might lead to impaired Block Design performance in WS comes from work by Bellugi and colleagues (e.g., Bellugi et al., 1988a; Bellugi, Sabo, & Vaid, 1988b;
Bellugi, Wang, & Jernigan, 1994) who reported that individuals with WS were able to choose the correct block faces, but that their solutions were not properly integrated into a global whole. A control group of individuals with Down syndrome (DS) showed the opposite pattern, namely greater global than local accuracy. However, individuals with DS also show an atypical profile of cognitive abilities (Klein & Mervis, 1999), and one must therefore be cautious when determining the extent to which group differences can be accounted for by deviance in the WS, as opposed to the DS, group. Nevertheless, this pattern of performance in WS is mirrored in drawing tasks, when compared to typically developing controls (Bertrand, Mervis, & Eisenberg, 1997).

To fully understand the nature of any deficit in Block Design performance in WS, studies need to isolate the different processing stages, segmentation and integration. A local processing bias at the level of perceptual segmentation has been reported in autism by Shah and Frith (1993). When presented with an adapted Block Design task, in which the model image was pre-segmented into the individual block faces, individuals with autism showed no or little effect of segmentation, compared to typically developing controls whose performance was facilitated by segmentation. This suggests that the group with autism perceive the model image more as a collection of parts than as a complete image. Following this approach, Farran et al. (2001) investigated the effects of segmentation in WS using a ‘Squares construction task’, a 2D version of the Block Design task (see Figure 1). Participants were given 4 squares, which resembled the block faces of the Block Design task but which were divided through the centre, either diagonally (oblique), or along the vertical/horizontal axis (nonoblique). They were then shown a model image, which they were asked to copy by placing the squares in a 2 by 2 formation within a square frame. The
results showed that the WS group benefited from segmentation to the same extent as typically developing controls. This suggests that individual with WS do not have a local bias at perception. Pani et al. (1999) provided further support for this suggestion using a visual search paradigm. They showed that the relative effects on search efficiency of the global configuration of the visual array, and the number of local elements it contained, did not differ between a group of individuals with WS and a group of typical adults. It appears then, that the poor performance on the Block Design task reported in WS does not relate to the perceptual stage of segmentation. A local bias in WS is more evident in production than perception tasks, suggesting that individuals with WS may suffer more from problems of integration, than of segmentation. Indeed, the studies by Bellugi and colleagues support this notion; in the Block Design task, their WS group showed difficulty in integrating the individual blocks into a coherent whole (e.g., Bellugi et al. 1988b). The impairments in drawing ability in WS also indicate difficulty with integration (e.g. Bertrand, et al. 1997).

Despite the progress made by considering the processing stages of the Block Design task in isolation, this task is not a pure measure of local and global processing; it draws upon a number of cognitive abilities. These include the ability to discriminate between the block faces (different patterns and different orientations of the same pattern) and the ability to plan a construction using verbal mediation or mental imagery. It is therefore possible that poor performance on block construction tasks in WS reflects these other abilities.

One of the factors above, verbal mediation, was investigated by Farran, Jarrold, and Gathercole (1999). We examined the possibility that verbal mediation is used to facilitate task completion in WS by measuring the relative contribution of verbal and non-verbal intelligence to the level of ability shown by individuals with
WS on the Performance subtests of the WISC. The results showed that the variance uniquely associated with verbal ability was significantly correlated with those tasks in which performance was relatively elevated. This did not include the Block Design task, which suggests that although individuals with WS can employ a verbal strategy for some tasks with a positive effect, this is not the case for Block Design.

In addition to examining the effects of segmentation on Block Design performance, Farran et al. (2001) also explored the second factor noted above, mental imagery. In the Squares construction task, Farran et al. (2001) manipulated whether squares were divided by oblique or non-oblique lines; evidence from typical development shows that oblique lines are more difficult to discriminate between than nonoblique lines (Cecala & Garner, 1986). Farran et al. (2001) found that their WS group showed the same pattern of errors on the Squares construction task as typically developing (TD) controls matched for non-verbal ability, but performed at a lower level of accuracy overall. Both groups showed an effect of obliqueness; with significantly longer RTs and reduced accuracy on oblique compared to nonoblique trials. However, the effect on RTs was larger in the controls than the WS group. This might have reflected the mental and manual manipulation strategies available to each group. Mental and manual manipulation abilities were measured independently by Farran et al. (2001) using a mental and a manual rotation task. The TD controls performed well on both of these tasks, while the WS group showed high levels of performance on the manual, but not the mental rotation task. Farran et al. (2001) suggested that the TD controls used a mental rotation strategy to complete the relatively easier nonoblique trials, and a less advanced manual rotation technique for the harder, oblique trials in the Squares construction task. In contrast, the WS group,
who were unable to perform mental rotation, may have used a manual strategy throughout.

Although the difference in performance on the Squares construction task observed between WS and TD individuals might well reflect differential use of mental and manual rotation strategies in preparing to construct the stimuli, perceptual differences in the groups’ ability to discriminate between different block patterns and orientations might also contribute to this effect. The perception of orientation in WS has been addressed by employing the Benton Line Orientation task (Benton, Varney, & Hamsher, 1978) in which the participant is asked which of 11 lines (oriented 18 degrees apart) matches the orientation of two target lines. Individuals with WS have consistently been found to be at floor in their performance on this task (Bellugi et al., 1988a; Rossen, Klima, Bellugi, Bihrlle, & Jones, 1996; Wang, Doherty, Rourke, & Bellugi, 1995). Although this may indicate that orientation discrimination is poor in WS, it may also reflect the fact that the task is not pitched at the appropriate level to properly test individuals with WS (Farran & Jarrold, 2003). Stiers, Willekens, Borghgraef, Fryns, and Vandenbussche (2000) designed the Pre-school Judgement of line Orientation task (PJLO), which has fewer lines in the display set than the Benton task. They found that the level of ability shown by individuals with WS on this task was not significantly different from their overall level of non-verbal ability, measured by the nonverbal subtests of the WPSSI (Wechsler, 1989). These mixed findings leave open the question of whether poor performance on the Squares construction might be attributed to problems of orientation perception.

Consequently, to further our understanding of the pattern of performance observed in the Squares construction task, the first experiment presented here directly examines the perceptual processing of oblique and nonoblique lines in WS. The
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second experiment then further examines one aspect of mental imagery that could also contribute to performance on Block Design tasks, namely the ability to transform a mental representation.

Experiment 1

Method

Participants

Twenty-two individuals with WS and 22 typically developing children participated in this experiment. This study took place approximately 8 months after the previous study in which 21 of the WS group in this study had taken part (Farran et al., 2001). The TD group were a different group of children, but recruited from the same school as that used in the Farran et al. (2001) study. The two groups were matched individually by performance on the RCPM (Raven, 1993), a measure of fluid intelligence. This task is a non-verbal perceptual task in which participants are shown a pattern or sequence in which a piece is missing. They are asked to choose which of six choice pieces is the missing piece. Some of the distracter choices included are reflections, orientations, and enlargements of the target, which raises the possibility of matching away any differences in performance. However, as these distracter choices occur infrequently, group differences in performance on experimental tasks should not be masked by this matching procedure. Participant details are shown in Table 1.

Six members of the WS group had received a “fluorescence in situ hybridisation” (FISH) test. This is a diagnostic test which checks for a deletion of the elastin gene on the long arm of chromosome 7 which occurs in approximately 95% of individuals with WS (Lenhoff, Wang, Greenberg, & Bellugi, 1997). All 6 cases received positive FISH results, thus confirming a deletion of the elastin gene. None of the WS participants had received negative FISH results. The remaining sixteen WS
participants had been diagnosed by medical practitioners before the WS genotype had been identified. This diagnosis was based on their unique cognitive, behavioural and facial characteristics. All WS participants had been recruited for a previous study from the records of the Williams Syndrome Foundation UK.

Table 1 about here

Design and Procedure

An A5 card was placed length-ways in front of the participant. This depicted the 4 target stimuli; 4 black and white squares divided through the centre, each displayed in one of 4 possible orientations, as shown in Figures 2a and b. In condition 1 the squares were divided by a diagonal line (obliques) and in condition 2 they were divided by horizontal/vertical lines (nonobliques). A sliding arrow was attached to the base of card, which was used to point to each of the four targets. The participant was shown a model image which was made up of the four possible orientations of stimuli (i.e. the 4 target squares) in a 2 by 2 formation, and example of which is illustrated in Figure 3 below. Red lines separated the image into the four quadrants. Participants were asked to match one of the target squares on their card, as indicated by the red arrow, to one of the 4 quadrants of the stimulus pattern by outlining it with their finger. The arrow indicated which square to identify sequentially, moving from left to right across the four possible targets. This sequence was repeated 4 times, thus there were 16 trials. The position of each of the 4 target squares in the model image appeared in each of the 4 quadrants once. Presentation of the model images was in a fixed random order, so that the correct position of the target in the image presented could not be predicted. Reaction time (RT) was measured using a stop watch. RT was
taken for all responses, but those for erroneous responses were not used. The number of correct responses was also recorded and treated as a second dependent variable.

**Results**

Group matching was analysed using a one-way ANOVA with RCPM score as the dependent variable and group as the independent factor (2 levels: WS, TD). This revealed that the groups were adequately matched: $F(1, 43) = 0.12, p = .74$. Data from the Squares discrimination task were analysed in two ways in terms of both the number of correct responses and response time (RT). Each analysis employed a 2 factor ANOVA with obliqueness as the within participant factor (2 levels: oblique, nonoblique), and group as the between participant factor (2 levels: WS, TD).

Preliminary analysis of the RT and correct response data was carried out using the Kolmogorov-Smirnoff one-sample test. This indicated that the data set was approximately normal and not in need of transformation prior to further analyses.

**Correct responses**

The number of correct responses across each condition are shown in Table 2 below. Analysis showed a significant main effect of group, $F(1, 42) = 4.79, p = .03$, partial $\eta^2 = .10$, due to less accuracy in the WS group than the TD controls. There was also a significant main effect of obliqueness, $F(1, 42) = 58.01, p < .001$, partial $\eta^2 = .05$, which reflected reduced accuracy on the oblique trials. The interaction between obliqueness and group was not significant, $F(1, 42) = 2.39, p = .13$, partial $\eta^2 = .05$.

**Response Time**

Response times for those trials in which the participant had given an incorrect response were replaced by a response time of 4.10 seconds, as this was the mean
correct response time across the groups and conditions. This is a conservative method of score adjustment. Mean RT by group and condition is illustrated in Figure 4. The analysis showed a main effect of obliqueness, $F(1, 42)=29.08, p<.001$, partial $\eta^2 = .41$, which was caused by slower responses to the oblique trials. The effect of group was not significant, $F(1, 42)=1.48, p=.23$, partial $\eta^2 = .04$. However, there was a significant group by obliqueness interaction, $F(1, 42)=4.20, p=.05$, partial $\eta^2 = .09$.

Analysis by groups indicated a significant effect of obliqueness in both groups; this was larger in the TD group, $t(21)=4.92, p<.001$, than in the WS group, $t(21)=2.55, p=.02$. Response times of the 2 groups to the nonoblique trials were very similar, $t(21)=.06, p=.96$, in comparison to a group difference which approached significance in responses to the oblique trials, $t(21)=-1.78, p=.09$.

**Z-score analysis**

Having established that the performance of the WS group on the Squares discrimination task differed from that observed in typical development, we were interested in how this difference compared to the level of performance measured in WS on the Squares construction task employed by Farran et al. (2001). A direct comparison of the absolute levels of performance of individuals with WS between these two tasks is not appropriate as one would not be able to ascertain the extent to which any differences in the absolute level of ability reflected impaired or unimpaired performance. Therefore, the two original data sets were transformed into z-scores on the basis of the performance of controls as this accounts for any difference in the relative difficulty of the two tasks which occur in typical development. In the present study, the correct response scores of the WS group were standardised separately for each task, based on the distribution of performance of each respective control group.
The two control groups in question had the same levels of nonverbal ability as measured by the RCPM. The mean RCPM score for the control group who completed the Squares construction task was 18.14 (S.D. =5.20), whilst the mean RCPM score for the control group who were given the Squares discrimination task was 18.18 (S.D.=4.89). This was confirmed statistically though independent t-tests which showed no significant difference between the scores of the two control groups (t(41)=.025, p=.98).

One participant with WS had taken part in the Squares discrimination task, but not the Squares construction task, hence they were excluded from this analysis. This left data from each of 21 individuals with WS on the two tasks. Descriptive statistics revealed similar levels of performance across the tasks in WS; Squares construction task: mean = −1.09, S.D.= 1.46; Squares discrimination task: mean= −0.79, S.D.= 1.19. These z-scores for WS performance on the two tasks were compared using a paired comparison t-test. This showed that the relative level of impairment of the WS group on the two tasks did not differ significantly; t(20)=0.84, p=.41.

**Discussion**

The data from the Squares discrimination task indicate that, in terms of the number of correct responses, individuals with WS were poorer than the TD controls, but showed the same pattern of performance, i.e., poorer performance on the oblique than the nonoblique trials. However, the RT analysis showed a variation between the groups. Both groups took significantly longer to respond in the oblique trials than the nonoblique trials, with a larger effect of obliqueness in the control group than in the WS group.

In both these data and those of Farran et al. (2001) there was an interaction between group and obliqueness in reaction times, but not in correct responses. We
(Farran et al., 2001) suggested that this might be attributable to differential strategies used during the processes involved in construction. However, the data from the Squares discrimination task demonstrate that this differential effect of obliqueness in RT is due to perceptual differences in processing between the groups. This is an important finding as it suggests that the difficulty in completing Block Construction tasks in WS is not purely constructional, but also relates to the perceptual processing of orientation. The group difference could be explained in two ways; the TD controls could be showing greater problems on oblique trials, or the WS group could be showing less facilitation for nonoblique relations. As the WS group are responding faster than the controls on oblique trials, the former explanation seems to best fit the data. The pattern of performance in the WS group therefore, is not necessarily a deficit, but it does indicate a deviance from typical development. However, the WS group were significantly less accurate than the controls in discriminating between stimuli in both oblique and nonoblique trials. This indicates that, in addition to a deviant processing style, there is a general impairment in orientation discrimination in this population.

The Z-score comparison of the performance of the WS group relative to the controls, across construction and discrimination versions of the Squares task, indicates that the additional demand of construction does not affect the relative level of impairment in WS. This direct comparison between the tasks provides evidence against the hypothesis that visuo-spatial construction/integration is particularly poor in WS. However, it is possible that this similarity in the level of impairment reflects the fact that on the Squares construction task accuracy was only affected by errors at the local level; global accuracy was constrained as the individuals were given a frame in which to place the squares. The results of previous investigations (e.g., Bellugi et
al., 1988b) suggest that the predominant cause of poor visuo-spatial construction abilities in WS in tasks such as the Block Design task is reduced accuracy at the global level, i.e. in placing the local elements together coherently. Facilitation at the global level provided by the frame in the Squares construction task, could explain why the level of performance on the two version of the Squares task did not differ significantly in WS.

Experiment 2

The findings from Experiment 1 suggest that individuals with WS are poor at orientation discrimination overall, and show a reduced effect of obliqueness on RT. These results imply that any tasks that present stimuli at differing orientations may have a detrimental impact on performance in WS. One such task is mental rotation (Farran et al., 2001); poor performance on this task in WS could reflect difficulty in orientation processing rather than, or in addition to, difficulties in mental image transformation as was first supposed (Farran et al., 2001). A problem with orientation coding could also contribute to the previously observed difference between mental and manual rotation tasks. Poor orientation discrimination is more likely to affect mental than manual rotation ability. This is due to the opportunity to make visual matches in a manual rotation task, which lessens the general load on orientation coding.

In order to test whether individuals with WS have a problem in mental transformation that is not contaminated by poor orientation coding, Experiment 2 examined size transformation abilities. Individuals were shown two different sized objects simultaneously and asked whether they are the same or different, regardless of any difference in size. Unlike mental rotation, this task does not involve orientation perception. Importantly, the task demands of size transformation are very similar to
those involved in mental rotation. First, neither task has a memory load, which could confound performance in this population (Jarrold, Baddeley, & Hewes, 1998b). Second, both tasks involve mental object manipulation, one by enlarging/ reducing the stimuli, the other by rotating the stimuli (Kosslyn & Koenig, 1992). Third, there is a linear relationship between response time and the transformation process in both of these tasks (Bundesen & Larsen, 1975; Shepard & Metzler, 1971), which provides evidence for mental transformation in both tasks. The similarity between mental rotation and size transformation tasks is also supported by studies investigating brain activation. Larsen et al. (2000), using Positron Emission Tomography (PET), found that dorsal areas were active during a size transformation task, similar to the activation seen in mental rotation.

This investigation has implications for the dorsal stream deficit hypothesis. The dorsal visual stream may support information for both mental image transformation (Alivisatos & Petrides, 1997; Larsen et al., 2001) and actions to objects (mental and manual; Wohlschlager & Wohlschlager, 1998). If poor mental rotation ability in WS (Farran et al., 2001) is due to a weak ability to perform image transformations, one should also observe poor performance on a size transformation task. However, if performance on the mental rotation task was influenced by the ability to discriminate between orientations in WS, then performance on the size transformation task should be comparatively better. This would indicate a fractionation in dorsal stream processing in WS.

**Method**

**Participants**

The testing session for Experiment 2 took place approximately 10 months after the testing session for Experiment 1. Two participant groups were employed; 21
of the original WS group employed in Experiment 1, and 21 typically developing (TD) individuals. As in Experiment 1, the TD controls were matched individually by score on the RCPM (Raven, 1993), and were recruited from the same school employed to recruit controls in Experiment 1. Participant details are shown in table 3.

Table 3 about here

Design and Procedure

Individuals were presented with an A4 booklet containing 40 stimulus pairs of abstract shapes, one pair on each page. Each pair of stimuli was presented in a landscape format, with the page divided horizontally down the middle by a black line. There was an abstract shape on each half of the page presented in a square box, as shown in Figure 6. Stimulus pairs differed in size of a ratio of 1:1 (no difference), 1:2, 1:3, 1:4, or 1:5. There were 4 target shapes each with 10 edges, one of which always appeared on the left of the page, as the smaller of the two presented shapes.

Individuals were instructed to say whether the shape on the right was the same or different from the target shape on the left, regardless of differences in size. Differences between the two shapes were subtle. Different shapes were constructed by changing 2 of the 10 lines only, by varying the angles between these 2 lines, and the adjacent lines, thus the whole shape had to be inspected before a response could be made. For each target shape, there were 5 same trials, one at each size ratio, and 5 different trials, one at each size ratio. Thus there were 40 trials, 10 for each target shape, with five different and five same pairs. Correct responses were recorded.

Response times were recorded from the moment the participant was shown the stimulus pair, until they produced a verbal response, using a stop watch.

Figure 5 about here
Results

A one-way ANOVA, with RCPM score as the dependent variable, and group as the independent variable (2 levels: WS, TD) revealed that the group matching was satisfactory: $F(1, 41)=.08, p=.79$. Data for the size transformation task were analysed in terms of the number of correct responses and in terms of response time (RT) for correct responses only. Preliminary analysis of correct response and RT data, using the Kolmogrov-Smirnov one-sample test, showed that response distributions were approximately normal and not in need of transformation prior to further analyses. ANOVA results are reported in terms of the between and within participant main effects for the factors of group and response type respectively, and in terms of within participant linear contrasts for the within participant factor of size ratio. Within participant contrasts are of interest due to previous evidence demonstrating that size ratio has a linear effect on speed and accuracy.

Correct responses

Figure 6 illustrates the level of performance of the two groups at each size ratio. For this analysis, the correct response data was partitioned into the responses to ‘same’ and ‘different’ trials. A 3 way ANOVA was employed on the number of correct responses with group (2 levels: WS, TD) as the between participant factor, and size ratio (5 levels: 1:1, 1:2, 1:3, 1:4, 1:5) and response type (2 levels: same, different) as the within participant factors. Descriptive statistics of these data are shown below in Table 4. The results showed that there was no significant main effect of group, $F(1, 40)=1.14, p=.29$, partial $\eta^2=.03$. There was a significant main effect of size ratio, reported as a linear trend: $F(1, 40)=40.39, p<.001$, partial $\eta^2 =.50$, which indicates that the number of correct responses decreased linearly as size ratio increased. The interaction between size ratio and group was not significant ($F<1$). The main effect of
response type was significant, $F(1, 40)=19.09$, $p<.001$, partial $\eta^2=.32$, due to increased accuracy on the ‘different’ responses. There was also a significant interaction between size ratio and response type, $F(4, 160)=5.85$, $p<.001$, partial $\eta^2=.13$, which is investigated below. The interaction between response type and group was not significant, $F(1, 40)=2.43$, $p=.13$, partial $\eta^2=.06$, and the 3-way interaction between response type, size ratio, and group was also not significant, $F(4, 160)=1.14$, $p=.34$, partial $\eta^2=.03$.

To investigate the interaction between ratio and response type, two one-way ANOVAs were employed, one for each response type, with size ratio as a within participant factor (5 levels). Analysis of the ‘same’ responses showed a significant main effect of ratio for ‘same’ responses, reported in terms of the linear contrast, $F(1, 41)=44.28$, $p<.001$, partial $\eta^2=.52$. This contrasted to the results of the analysis of the ‘different’ response, where the main effect of size ratio was not significant, reported in terms of the linear contrast, $F<1$.

Response times
Response times were averaged for each participant across the 4 target shapes. A three way ANOVA was carried out with one between participant factor of group (2 levels: WS, TD) and two within participant factors: size ratio (5 levels: 1:1, 1:2, 1:3, 1:4, 1:5), and response type (2 levels: same, different). Empty cells were replaced by the mean RT, across both of the groups, for responses to the largest (1:5), and arguably the hardest size ratio. As incorrect responses reflect a high level of difficulty, this value most closely represents performance on these trials. Due to the small number of empty cells replaced (<1%), the reduction in variance was minimal.
The ANOVA showed a nonsignificant effect of group, $F(1,40)=1.65, p=.21$, partial $\eta^2=.04$. There was a main effect of size ratio, reported here in terms of a significant within participant linear contrast, $F(1, 40)=6.93, p<.05$, partial $\eta^2 = .15$ which indicates that RT increased linearly with increasing size ratio. The main effect of response type approached significance, $F(1,40)=3.14, p=.08$, partial $\eta^2 = .07$, with longer RT on the different trials. Importantly, there was no group by size ratio interaction, $F<1$, indicating that both groups were affected by size ratio in the same way. There were no significant 2-way or 3-way interactions between the response type factor and the two remaining factors (response type by group: $F<1$; size ratio by response type: $F(4, 160)=1.34, p=.26$, partial $\eta^2 = .03$; size ratio by response type by group: $F<1$). These data are illustrated, averaged across response type, in Figure 7.

Z-score analysis

A z-score analysis was carried out to compare the relative level of ability of the WS group on the mental rotation task employed in our previous study (Farran et al., 2001) to that achieved on the size transformation task in the present study. The control groups employed here and in Farran et al. (2001) had comparable levels of nonverbal ability as measured by the RCPM. Mean RCPM score for control group who completed the previous mental rotation task was 18.14 (S.D.= 5.20). Mean RCPM score for the controls who were given the current size transformation task was 17.57 (S.D.= 5.00). An independent $t$-test revealed that the RCPM scores of the two control groups did not differ significantly ($t(42)=0.01, p=.93$) and thus that they had comparable levels of nonverbal ability. Thus, the two original data sets of the WS
group were transformed into z-scores to give two standardised measures of performance based on the distribution of scores of the appropriate group of controls.

One participant with WS had taken part in the mental rotation task, but not the size transformation task, and similarly one individual had been included in the size transformation task, but not the mental rotation task. The data from these two individuals were excluded from the analysis. The data remaining was from 20 individuals with WS on each of the mental imagery tasks. Mean z-scores for performance on the two mental image transformation tasks were: mental rotation mean = -1.00, S.D. = 0.667; and size transformation mean = -0.255, S.D. = 0.667. To compare WS performance across the two tasks, the z-scores for the two tasks were subjected to a paired comparison t-test. This demonstrated that level of performance of the WS group on the size transformation task was significantly superior to that on the mental rotation task, t(19) = 4.36, p < .001.

Discussion

The results of Experiment 2 indicate that individuals with WS behaved in a comparable way to TD controls on a size transformation task. For both groups there was a linear effect of the size difference on performance as found in previous investigations (Besner, 1983; Bundesen & Larsen, 1975; Larsen & Bundesen, 1978). This is an important finding as it indicates not only that individuals with WS can perform mental image transformation when assessed using a size transformation task, but also that they can complete the task to the same level as their typically developing non-verbal matched peers. One can therefore assume that this ability is available to individuals with WS when completing a block construction task, and does not contribute to their particularly poor level of performance on such tasks. In turn, this suggests that not all aspects of dorsal stream functioning are equally impaired in WS.
A difference between same and different response patterns is not an uncommon finding in the literature on size transformation (e.g., Besner, 1983; Besner & Coltheart, 1976; Kubovy & Podgorny, 1981). We also found a linear decrease in accuracy with increasing size ratio from responses to the ‘same’ trials only. Responses to the ‘different’ trials were not affected by increasing size ratio. This difference in response patterns for same and different trials suggests that the increased difficulty experienced with increasing size ratio, must relate to the act of size transformation itself, rather than a general difficulty caused by an overall increase in cognitive demands. We suggest that this difference in response patterns is best explained by response uncertainty, which could cause participants to adopt strategic response patterns. The pattern of performance in the present study appears to indicate that as trials became more difficult, individuals became less able to perform the size transformation accurately and, when unsure defaulted to a ‘different’ response. On the ‘same’ trials, this default response was incorrect, which can explain why accuracy decreased with increasing size ratio and why, on ‘different’ trials accuracy did not decrease with increasing size ratio, but stayed constant across the five size ratios (see Besner, 1983; and Jolicoeur & Besner, 1987 for alternative accounts).

The results of Experiment 2 contrast sharply with those in our previous study (Farran et al., 2001) where level of performance of the group of individuals with WS on a mental rotation task, also a measure of mental image transformation, was significantly poorer than that of a control group who were again matched by score on the RCPM. The Z-score analysis demonstrated a significant difference in the level of ability of the individuals with WS on the two tasks, relative to matched controls; performance was superior on the size transformation task in WS than on the mental rotation task. It appears that the ability to mentally transformation images is available
to individuals with WS, and that the poor level of mental rotation ability in WS can be accounted for by the orientation perception requirements of this task. The orientation requirements of mental and manual rotation tasks are thought to be properties of the dorsal visual stream, (Wohlschlager & Wohlschlager, 1998), as is the ability to transform mental images (Cohen et al., 1986; Larsen et al., 2000). These results suggest that dorsal stream functioning is fractionated in WS; there is a relative deficit in some, but not all, aspects of dorsal stream functioning.

General Discussion

The present study provides a more precise illustration of the deficits in visuo-spatial ability in the WS population. It is well known that performance on block construction tasks in WS is particularly poor (Mervis et al., 1999). Until now, the main focus of investigation into this deficit has been on the local and global processing requirements of the task. Although fruitful, this approach has ignored many other factors involved in block construction. Experiment 1 elaborated on the hypothesis that there is a local bias in production tasks in WS (Bellugi et al., 1988; Farran & Jarrold, 2003). The results indicated that poor block construction performance in WS is not necessarily due to weak constructional abilities, but also due to atypical perception, in particular unusual orientation processing.

Experiment 2 examined the dorsal stream deficit hypothesis (Atkinson et al., 1997) by investigating whether size transformation was equally impaired as mental rotation among individuals with WS. The level of performance of the WS group on the size transformation task did not differ from that of TD controls. This in turn suggests that the weak mental rotation ability previously reported was due to a difficulty in processing orientation, rather than performing mental transformation. Both mental rotation and the dorsal stream version of the post box task employed by
Atkinson et al. (1997) involve processing orientation. We suggest that it is the orientation processing properties of these tasks that lead to impaired performance, rather than a general deficit in dorsal stream processing.

Our conclusion that orientation discrimination is impaired in WS appears to contradict the findings of Stiers et al. (2000) who found that the ability to identify a target line amongst distracters may be no poorer than overall non-verbal level of ability in WS. However, this difference in results could reflect the choice of targets and distracters. In the Squares tasks, orientations are perpendicular to one another, or are the same. This may lead to greater demands being placed on the orientation discrimination system than in a task involving comparison of line orientations.

In addition to the implications that the present results have for understanding WS, this study also adds to the literature regarding the fractionation of mental imagery into separate subsystems. In particular, the data question whether all mental image transformations are served by one subsystem (cf. Kosslyn & Koenig, 1992), or at least question whether other factors (such as orientation discrimination) contribute more to mental rotation than size transformation.

Individuals with WS show an overall impairment in visuo-spatial cognition. Even on the tasks mentioned here where performance is at the level of controls, the control groups are many chronological years younger than the WS group. However, this deficit in visuo-spatial abilities appears to comprise a mixture of simple delay on some tasks, but also of a pattern of deviant performance on others. Indeed, the present study has revealed two important findings. First, individuals with WS do not have typical perceptual abilities – they show deviant orientation processing abilities. It appears then, that a local bias in construction is not the only contributor to poor visuo-constructive abilities in WS. Second, dorsal stream functioning in individuals with
WS appears to be fractionated. This population are able to use image transformation for mental size transformation, but are relatively poorer in completing a mental rotation task.

If a number of factors constrain performance on a task, one must necessarily consider the relative importance of these factors and the interaction between them in determining any delay in performance seen in an atypical group. This study demonstrates that a detailed analysis of the factors involved in task completion, can result in a precise understanding of the underlying deficits that give rise to a specific and deviant processing style.
References


Table 1: Participant details

<table>
<thead>
<tr>
<th>Group</th>
<th>RCPM: Mean(SD)</th>
<th>CA(years; months): mean(SD)</th>
<th>Range (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WS (N=22)</td>
<td>17.68(4.89)</td>
<td>21;3 (8;8)</td>
<td>10;2-39;2</td>
</tr>
<tr>
<td>TD (N=22)</td>
<td>18.18 (4.89)</td>
<td>6;6 (1;6)</td>
<td>5;9-7;9</td>
</tr>
</tbody>
</table>

Note: scores of 17 and 18 on the RCPM represent the 50% percentile for a typical child of 6;6 and 7;0 years respectively (Raven, Raven & Court, 1998)
Table 2: Correct responses on the Squares discrimination task

<table>
<thead>
<tr>
<th>Group</th>
<th>Nonoblique: mean (SD)</th>
<th>Oblique: mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>WS</td>
<td>10.91(3.87)</td>
<td>8.41(3.66)</td>
</tr>
<tr>
<td>TD</td>
<td>13.68(2.30)</td>
<td>9.91(3.98)</td>
</tr>
</tbody>
</table>
Table 3: Participant details

<table>
<thead>
<tr>
<th>Group</th>
<th>RCPM: mean (SD)</th>
<th>CA(years; months):</th>
<th>Range (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>mean(SD)</td>
<td></td>
</tr>
<tr>
<td>WS (N=21)</td>
<td>18.00(5.13)</td>
<td>21;2(7;10)</td>
<td>11;0-33;4</td>
</tr>
<tr>
<td>TD (N=21)</td>
<td>17.57(5.00)</td>
<td>6;3(0;6)</td>
<td>5;5-7;4</td>
</tr>
</tbody>
</table>

Note: scores of 17 and 18 on the RCPM represent the 50% percentile for a typical child of 6;6 and 7;0 years respectively (Raven, Raven & Court, 1998)
Table 4: Correct responses on the size transformation task

<table>
<thead>
<tr>
<th>Group</th>
<th>Size ratio: mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1:1</td>
</tr>
<tr>
<td>WS</td>
<td>6.67(1.20)</td>
</tr>
<tr>
<td>TD</td>
<td>7.10(1.09)</td>
</tr>
</tbody>
</table>
Author Note

This research was supported by a research studentship awarded to the first author, from the Williams Syndrome Foundation of the United Kingdom. We would like to thank those members of the WSF who have kindly participated in this study and the staff and students of Barton Hill Infant and Nursery School for their cooperation with this work. We would also like to thank Axel Larsen for helpful comments regarding the results of Experiment 2. 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. Squares construction Task: Nonsegmented and Segmented Versions of Oblique and Nonoblique Patterns. The Design is to be Constructed from Four Identical Squares; Oblique or Nonoblique.

Figure 2a. Target stimuli for the Oblique trials in the Squares discrimination task

Figure 2b. Target stimuli for the Nonoblique trials in the Squares discrimination task

Figure 3. Example model images for the nonoblique trials and the oblique trials

Figure 4. Group by obliqueness interaction on the Squares discrimination task

Figure 5. Example of stimulus pairs (size ratio: 1:1; response type: ‘different’).

Figure 6. Number of correct responses made on the size transformation task

Figure 7. Response times to correct responses on the size transformation task
Figure reprinted by permission from *Journal of Child Psychology and Psychiatry*, 2001, 42, 719-728.
The graph shows the reaction time (RT in seconds) as a function of the ratio of 1:x. The data is represented for two conditions: WS and TD. The x-axis represents the ratio of 1:x, while the y-axis represents the reaction time in seconds. The error bars indicate the variability in the data.