RUNNING HEAD: Location memory and Williams syndrome

Strategies and Biases in Location Memory in Williams Syndrome

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Abstract

Individuals with Williams syndrome (WS) demonstrate impaired visuo-spatial abilities in comparison to their level of verbal ability. In particular, visuo-spatial construction is an area of relative weakness. It has been hypothesised that poor or atypical location coding abilities contribute strongly to the impaired abilities observed on construction and drawing tasks (Farran & Jarrold, 2005; Hoffman, Landau & Pagani, 2003). The current experiment investigated location memory in WS. Specifically, the precision of remembered locations was measured as well as the biases and strategies that were involved in remembering those locations. A developmental trajectory approach was employed; WS performance was assessed relative to the performance of typically developing (TD) children ranging from 4- to 8-years-old. Results showed differential strategy use in the WS and TD groups. WS performance was most similar to the level of a TD 4-year-old and was additionally impaired by the addition of physical category boundaries. Despite their low level of ability, the WS group produced a pattern of biases in performance which pointed towards evidence of a subdivision effect, as observed in TD older children and adults. In contrast, the TD children showed a different pattern of biases, which appears to be explained by a normalisation strategy. In summary, individuals with WS do not process locations in a typical manner. This may have a negative impact on their visuo-spatial construction and drawing abilities.

Strategies and Biases in Location Memory in William Syndrome

Introduction

Williams syndrome (WS), a rare genetic disorder, occurs in approximately 1 in 20, 000 live births (Morris & Mervis, 1999). Individuals with WS show an unusual cognitive profile in which verbal abilities are superior to visuo-spatial abilities (e.g. Udwin & Yule, 1991). Furthermore, an atypical pattern of strengths and weaknesses can be observed within each domain (Karmiloff-Smith et al., 1997; Farran & Jarrold, 2003). The visuo-spatial domain is characterised by relative strengths in face processing and perceptual identification, and weaknesses in drawing and construction tasks (Karmiloff-Smith, 1997; Farran, Jarrold & Gathercole, 2003).

The ability to represent an object's location appears to be a relative weakness in WS. Hoffman, Landau, & Pagani (2003) suggest that poor performance on block construction tasks, a hallmark of WS (Mervis, 1999), can be explained by deficits in coding the identity and location of each component part. Hoffman et al. (2003) isolated these two task demands; in an identity task and a location task, participants were shown a model image with one block cued. In the identity task, they then chose an identity match from a set of 2D block faces. In the location task, participants were shown a single block, and a copy space of possible block locations, and asked to place the block in the correct location in the copy area. WS performance was significantly poorer than control participants on both tasks (comparisons across tasks were not made). Paul, Stiles, Passarotti, Bavar and Bellugi (2002) also employed a location matching task, and similarly report poor abilities in WS. However, in both studies, controls were matched by overall mental age, which necessarily assumes a group difference in visuo-spatial cognition. Further investigation is required to determine how performance on such tasks relates to the visuo-spatial profile in WS.

Farran and Jarrold (2005) present further evidence for atypical location coding in WS. They investigated two types of spatial relations, using tasks adapted from Koenig, Reiss and Kosslyn (1990). Coordinate spatial relations refer to the encoding of fine grain information, e.g. precise locations, specific distances. In this task participants judged whether a ball was ‘in’ or ‘out’ (within or beyond a certain distance from a bat). Categorical spatial relations are regions of space that cover a range of values (see Kosslyn & Koenig, 1992). In this task, individuals classified a ball as ‘above’ or ‘below’ a bat. The WS group performed at a comparable level to TD children matched by visuo-spatial ability. However, on both tasks, the WS group unexpectedly showed response biases in the opposite direction to TD children and adults. This suggests that individuals with WS categorise spatial locations in an atypical manner. Although these alternative coding strategies did not negatively affect level of performance on the spatial relations tasks, the tasks were perceptual. It is therefore possible that poor performance on production tasks relates to a negative impact from such strategies.

Object location processing is thought to be a function of the dorsal visual stream (Ungerleider & Mishkin, 1982). Thus, impaired location coding in WS could inform the ‘dorsal stream vulnerability’ hypothesis, which explains that visuo-spatial cognition in WS can be accounted for by weaker dorsal than ventral stream functioning (e.g. Atkinson et al., 1997). Indeed, in support of this, studies of short term and long term memory have demonstrated a dissociation between impaired memory for spatial location, relative to visual identity in WS (e.g. Vicari et al., 2004; Vicari, Bellucci, & Carlesimo, 2006; Vicari, Bellucci, Santa Marinella, & Carlesimo, 2003). However, on account of mixed support (Atkinson et al., 2006; Jordan, Reiss, Hoffman & Landau, 2002), current thinking points towards a fractionation of dorsal functions in WS (see Meyer-Lindenberg et al., 2004). Location coding in WS is related to current hypotheses in the present experiment. Before introducing this Experiment, spatial location coding strategies in typical development are discussed.

The Category Adjustment model (Huttenlocher, Hedges & Duncan, 1991) details spatial location coding in typical adults and children. The model describes two steps (note that these are comparable to coordinate and categorical spatial relations; Kosslyn & Koenig, 1992). Location is first estimated using fine-grained information, i.e. the distance and direction of an object's location from an edge/ another object. These estimates are then adjusted using spatial regions / categories. Adjustment is typically towards a prototype at the category centre: the prototype effect. When fine-grained information is less certain, categorical information is more strongly weighted. In turn, the extent of influence from categorical information determines the strength of any bias in location coding. For example, individuals overestimate distances between objects that are in different regions and underestimate distances between objects that are in the same region, known as the subdivision effect (Huttenlocher et al., 1991).

Plumert and Hund (2001) investigated location coding in typical 7-, 9- and 11-year-old children and adults. Participants learnt twenty locations within a 32 $inch^2$ box. Boundary salience increased across three conditions: no boundaries; box divided into four quadrants by lines; or by walls. All groups showed increased accuracy with stronger boundary salience, and overestimated between quadrant distances. The 11-year-olds and adults showed significantly longer estimates for between than within quadrant distances, indicating a subdivision effect in the older groups. However, the prototype effect was not always evident; the adults showed no observable displacement (Experiment 1) or a prototype effect (Experiment 2), and the children displaced locations away from the centre of a region and towards the model corners. Thus, it appears that subdivision effects cannot always be explained by a prototype effect, and that these two effects are independent. Developmentally, it appears that category prototypes are not employed by younger children.

The present study investigated location coding in WS, using a task based on Plumert and Hund (2001). As individuals with WS have a poor level of visuo-spatial memory in WS (e.g., Jarrold, Baddeley & Hewes, 1998; Vicari, Belluci & Carlesimo, 2006; Vicari et al., 1996), a pilot study was carried out. This determined that participants should be asked to learn eight locations. The use of boundaries and prototypes for coding spatial location seems to be atypical in WS (Farran & Jarrold, 2005). The biases observed in the tasks described by Farran and Jarrold (2005) can speculatively be explained in relation to fewer (coordinate task) or different (categorical task) category boundaries imposed by the WS group than the controls, which results in the employment of different category prototypes. Category boundaries and prototypes will be investigated systematically in this study. The biases involved in location memory in WS may give some insight into the strategies that an individual employs to code a location, which in turn might go some way to explaining the poor drawing and construction abilities observed in this population.

Method

Participants

Fifteen individuals with WS were recruited from the records of the Williams syndrome foundation, UK. All individuals had received positive diagnosis of WS based on 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). Five groups of typically developing children also took part. There were ten individuals in each group; aged approximately 4, 5, 6, 7, and 8 years old. The level of visuo-spatial ability of all participants was assessed using the Ravens Coloured Progressive Matrices (RCPM; Raven, 1993), a recognised non-verbal measure of fluid intelligence (Woliver & Sacks, 1986). From previous experience, and the scores on the RCPM, it was estimated that the age range of the typically developing children was appropriate to cover the range of abilities on the experimental task exhibited by the WS group. Table 1 illustrates the RCPM raw scores, and chronological age of each group. This table also shows p-values for independent t-tests, which compared the WS group to each TD group on the RCPM. The WS group's RCPM spores were most similar to those of the 6- and 7-year-old groups.

Materials

The Experiment was based on that of Plumert and Hund (2001). An open square box was employed, which measured 32 inches square and 13 inches high. This was referred to as a 'house'. The participant's task was to remember the location of eight objects, which were placed in the house. The objects were wooden toys that were replicas of household objects as follows: washing machine, vacuum cleaner, armchair, table, television, standing lamp, sewing machine and shelf unit. Objects were of approximately similar sizes, and no larger than two inches in length or width.

There were four removable floors which could be put into the house. These slotted between the base of the house and a layer of clear Perspex. Two of the floors had eight black dots on them, less than 1/8th inch in diameter, and were used in the training trials. The eight dots indicated the to-be-remembered locations and were arranged so that there were two in each quadrant. Six of these objects also formed two location triads. The triads were such that two objects were in the same quadrant, and the third was in an adjacent quadrant. The middle object of each triad was 6 inches from the object in the same quadrant and 6 inches from the object in the adjacent quadrant. Thus, the triads created two sets of between quadrant and within quadrant distances between objects. Target locations are illustrated in Figures 1a and 1b. The third floor was a plain white floor, used in the testing trials. The fourth floor was a grid of x and y coordinates separated by half-inch intervals, used to measure location estimates.

There were two conditions: ‘no walls’ and ‘walls’. For the walls condition, the house was divided into quadrants (16 inches square) by opaque walls of the same height as the model.

Design and Procedure

Participants were tested on the no walls condition first as this enabled one to observe whether participants spontaneously imposed boundaries. This was followed by the walls condition, in which the quadrant boundaries were imposed by the solid walls. The two conditions were separated by two other short tasks, not reported here. This was to reduce any interference of object locations from one condition to the next. The training floors were counterbalanced across no walls and walls conditions.

With the training floor in place, participants watched the experimenter place the eight objects in the correct locations. They were instructed that the objects would be removed and that they would then be asked to place the objects in the correct locations themselves. When the participant indicated that they were ready, the experimenter removed the eight objects, randomised them, and placed them in a pile for the participant to draw from. The participant then placed the objects on the dots in the house. When the participant had placed all objects, the experimenter gave them the opportunity to make any changes. The experimenter recorded placements. They then corrected any errors whilst explaining these to the participant. Following this correction method, the training procedure was repeated twice more. If the participant showed 100% accuracy on the first two training trials, the third training trial was not administered.

After the training trials, participants took part in the test trial. The floor with dots was removed and replaced by the plain white floor. Participants were again asked to place the objects in the correct locations, this time using their memory of where the eight locations were. The white floor was then removed, and the scoring floor was inserted in order to measure the x and y coordinates of each of the eight object placements. The walls condition was identical to the no walls condition except that the walls dividing the space into quadrants were in place, and remained so through training and test trials.

Results

Four of the WS group did not complete the walls condition due to fatigue. One of these individuals completed the training trials but not the test trials of the walls condition. Thus, the WS group has an N of 11 (12 for training trial analysis).

Training trials

Individuals received a score out of eight for the number of correct object placements. This was recorded for each of the three training trials (and the test trial, reported later, see Table 2). High accuracy by the end of training ensures fewer redundant object placements at the test trial.

In order to determine the effectiveness of the training phase, the absolute level of performance for the WS group and each of the five TD control groups was compared to a ceiling score of eight. One-sample t-tests revealed that the WS group and the 4-year-old TD group never reached ceiling performance in either the walls or the no walls conditions ($p<.05$ for all). In the no walls condition, the remainder of the control group (5- to 8-year-olds and adults) showed ceiling performance throughout training trials 1 to 3 ($p>.05$ for all). In the walls condition, the 5-year-olds did not reach a ceiling level of performance ($p<.05$), the 6-year-olds reached ceiling by training trial 3 ($t(9)=-1.91$, $p=.09$) and the seven-year-olds reached ceiling at training trial 2 ($t(9)=-1.72$, $p=.12$). The eight-year-olds scored a maximum score of eight for all training trials in both conditions.

Optimally, one would anticipate that the absolute levels of ability would be close to ceiling at the end of the training trials. This was true of the majority of participants, thus the correct balance of reducing the possibility of fatigue, but optimising performance was reached.

Table 2 about here

Test trials: Location memory bias

Analysis of location memory bias was performed only on those object placements that were considered to be based on location memory, therefore random object placements (guesses) were excluded from analysis. As this experiment was not designed as a measure of object identity accuracy, placements classified as valid include both accurate object placements and transpositions (i.e. where two locations were accurate, but the objects had been swapped around) and are shown in Table 2. A second rater coded the correct object placements, transpositions and valid placements (the sum of correct object placements and transpositions) for performance on the wall and the no wall condition of 12 individuals (two participants from the WS group and the 4, 5, 6, 7, and 8-year-old TD groups). Inter-rater reliability analysis gave Cohen's Kappa = 0.61, which indicates substantial agreement (see Viera & Garrett, 2005). For the TD groups aged 5- to 8- years, the number correct did not differ significantly from a ceiling score of 8 ($p>.05$). For the WS group and TD 4-year-old group, performance was not quite at ceiling, but at a respectable level (WS, no wall: $t(10)=-2.63$, $p=.03$; WS, wall: $t(10)=-2.67$, $p=.02$; TD 4-year-old, no wall: $t(9)=-4.88$, $p=.001$, TD 4-year-old, wall, $t(9)=-4.12$, $p=.003$).

There were three dependent variables: exactness scores, between and within quadrant distance and centre of quadrant displacement. Where appropriate, the TD groups were combined into a single TD group to analyse developmental trajectories of the typical performance using ANCOVA, with chronological age (CA) as a covariate. WS performance was not consistently related to CA or RCPM score, and so could not be included in ANCOVA analyses. As such, WS performance was compared to each of the five TD groups and a ‘matched’ group was selected for analysis using ANOVA.

Exactness scores

Exactness scores measure displacement in inches between each observed object placement and the actual location. The exactness score for each individual, is a mean of their displacement distances, thus lower scores indicate higher levels of accuracy: a score of zero represents 100% accuracy.

ANCOVA was carried out on TD performance, with a within participant factor of wall condition (2 levels: no wall, wall) and CA as the covariate. Accuracy score was negatively related to CA, $F(1, 48)=22.89$, $p<.001$, partial $\eta^2 =.32$, and did not interact with wall condition, $F<1$. The main effect of wall condition was not significant, $F<1$. The mental age (MA) equivalent for the level of ability of each individual with WS was calculated by matching WS exactness scores to the typically developing trajectory of exactness scores. Any extrapolation beyond the age range of the TD data is based on the assumption that the trajectory remains linear, and so must be treated with caution. Nevertheless, this revealed first that there was a large range of abilities in the WS group, and second that MA for the WS groups was substantially lower in the wall condition (mean (S.D.): 20.20 (40.84) months) than in the no wall condition (mean (S.D.): 49.88 (29.73) months) (see Figure 2).

WS performance was compared to each TD group separately using independent t-tests. As this procedure was solely for group selection purposes, no corrections were made. This showed that the WS group performed at a level lower than the 6-, 7- and 8-year-olds for both wall and no wall conditions ($p<.05$ for all), below the 5-year-old level for the wall condition (*p*=.004), but not the no wall condition (*p*=.56), and at a similar level to the 4 year olds for both wall (*p*=.21) and no wall (*p*=.88) conditions. As such, the 4- and 5-year-old TD groups were combined for comparison against WS performance. ANOVA with a between participant factor of group (WS, TD 4 to 5-year-olds) and wall condition as a within participant factor was carried out. This showed no main effect of group, $F(1, 29)=3.49$, $p=.07$, partial $\eta^2=.11$ (marginal direction: WS<TD 4 to 5-year-olds) or wall condition, $F(1, 29)=1.36$, $p=.25$, partial $\eta^2=.05$. However, the interaction between group and wall condition was significant, $F(1, 29)=5.05$, $p=.03$, partial $\eta^2=.15$. This was due to poorer accuracy in the WS group than the TD 4 to 5-year-old group for the wall condition ($t(29)=2.67$, $p=.01$), but not the no wall condition ($t(29)=0.31$, $p=.76$). This differential pattern for the WS group and the TD children suggest different strategies for remembering locations, as is illustrated in Figure 3.

Figures 2 and 3 about here

Between and Within quadrant distance estimates

Due to guess responses, some object placements had been eliminated from the data. This affected the number of participants with a full set of between and within quadrant distance estimates. Those participants with missing data were excluded from this set of analyses: participant numbers were 10 individuals with WS and 46 TD participants. Between and within quadrant distances are illustrated in Figure 4. Between and within quadrant distances of 6 inches indicate 100% accuracy. Thus, observed distances below or above 6 inches indicate underestimates and overestimates of distance respectively. The distribution of the no wall condition, within and between quadrant distances were not normal for the TD group. Thus, logtransformed variables were employed for all analyses. Descriptive statistics showed that all estimate means were overestimates. However, significant overestimates were demonstrated for between distance estimates only in the WS group (no wall condition, between, p=.02; wall condition, between, p=.001; within quadrant distances, p>.05 for both). The TD group showed more consistent overestimates (no wall, between, p=.06; wall, between, p<.001; no wall within, p<.001; wall within, p<.001).

ANCOVA of TD performance was carried out with within participant factors of wall condition (no wall, wall) and distance estimate (between quadrant, within quadrant) and CA as the covariate. This demonstrated smaller biases with increased CA, $F(1, 44)=9.59$, $p=.003$, partial $\eta^2=.18$. The main effect of wall condition was not significant, $F<1$. Overall, between distance estimates were less biased than within distance estimates, $F(1, 44) = 12.21$, $p=.001$, partial $\eta^2=.22$, although a significant interaction ($F(1, 44) = 4.84$, $p=.03$, partial $\eta^2=.10$) revealed that this was driven by the wall condition only (wall condition: $F(1, 44)=12.80$, $p=.001$, partial $\eta^2=.23$; no wall condition: $F<1$). The relationship between performance and CA was differentiated across conditions such that CA was related to a reduced bias for both types of within quadrant estimates, but showed no relationship to between distance estimates (between, wall $F<1$; between, no wall, $F<1$; within, wall, $F(1, 44)=19.80$, $p<.001$, partial $\eta^2=.31$; within, no wall, $F(1, 44)=8.27$, $p=.01$, partial $\eta^2=.16$).

Independent t-tests revealed that the performance of the WS group was most similar to the 5- and 6-year-old groups (5- and 6-year-olds, all paired comparisons, p>.05), but also showed similarities to the other groups (4-, 7- and 8-year-olds, at least three out of four comparisons, p>.05). As such, all five TD groups were combined into a single TD group for comparison with the WS group. ANOVA was carried out with a between participant factor of group (WS, TD 4- to 8-year-olds) and within participants factors of wall condition and distance estimate. The main effect of group was not significant, $F(1, 54)=2.65$, $p=.11$, partial $\eta^2=.05$. The main effect of wall condition was significant, $F(1, 54)=13.59$, $p=.001$, partial $\eta^2=.21$ (bias: no wall<

wall). The main effect of distant estimate was not significant, $F(1, 54)=1.42$, $p=.24$, partial $\eta^2=.03$. However, distance estimate interacted with group ($F(1, 54) = 8.01$, $p=.01$, partial $\eta^2=.13$, such that the TD group were less biased for between than within distance estimates, $F(1, 45)=4.99$, $p=.03$, partial $\eta^2=.10$. The WS group showed the opposite pattern in terms of means, but this was not observed as a significant main effect of distance estimate, $F(1, 9) = 2.21$, $p=.17$, partial $\eta^2=.20$. Remaining interactions were not significant (wall condition by group, $F<1$; wall condition by distance estimate, $F(1, 54)=2.01$, $p=.16$, partial $\eta^2=.04$; wall condition by group by distance estimate, $F<1$).

Figure 4 about here

In summary, the TD group showed larger within than between quadrant distance estimates in the wall condition, which is the opposite pattern to that predicted by a subdivision effect. This differentiated them from the WS group who showed a bias to overestimate between quadrant distances only, which gives some indication of an opposing pattern, relative to TD performance. Although, note that this distance estimate difference was not significant, possibly due to lack of power. Across WS and TD groups, performance on the no wall condition showed less bias than on the wall condition. For the TD group, a reduced bias in distance estimates in the wall compared to the no wall condition was not predicted for each age group; this was supported by the lack of effect of wall condition when CA was covaried out.

Centre of quadrant displacement

The analysis of between and within quadrant distance estimates above, indicates that the TD group show some evidence of strategy use, which was different from the WS group. To determine whether the strategies used involved a prototype in the centre of a quadrant as suggested by the Category Adjustment model (Huttenlocher et al. 1991), displacement to the centre of quadrants was assessed. This was calculated by subtracting the actual distance from an object to the quadrant centre from the observed distance. Thus, negative scores and positive scores indicate a displacement towards or away from the quadrant centre respectively. Neither group showed a relationship between performance and CA or RCPM score, and so the TD groups are treated as a single TD group, and developmental trajectories are not analysed. Descriptive statistics show a significant displacement away from the quadrant centre in both the WS and TD groups in the no wall condition only (WS, wall condition, mean displacement = -0.06 p=.88; WS, no wall condition, mean displacement = 1.33, p=.001; TD, wall condition, mean displacement = 0.25, p=.12; TD, no wall condition, mean displacement = 0.88, p<.001). ANOVA with a between participant factor of group and a within participant factor of wall condition revealed no main effect of group, F<1. However, displacement was significantly stronger in the no wall condition than the wall condition, $F(1, 59)=23.81$, $p<.001$, partial $\eta^2=.29$. This did not interact with group, $F(1, 59)=3.27$, $p=.08$, partial $\eta^2=.05$. These results suggest that a category prototype is being employed in the no wall condition only. The bias is not towards, but away from a prototypical location, which is consistent with Plumert and Hund (2001).

Discussion

Location memory in WS was compared to five groups of TD children. Although not the primary focus, the training phase has comparative value to previous studies as it involved matching objects to locations. Hoffman et al. (2003) and Paul et al. (2002) report that location matching was poor in WS. The current results show a similar difficulty and, importantly, can qualify the finding; the ability to match locations in WS is similar to that of a typical 4-year-old, despite a general level of visuo-spatial cognition (measured by RCPM score) at the level of a 6- to 7-year-old. Thus, location memory represents a relative weakness within the visuo-spatial domain in WS. As this is a function of the dorsal visual stream, one could argue that this weakness supports a ‘dorsal stream vulnerability’ (Atkinson et al., 1997) in WS.

In the training phase, all participant groups found it easier to match an object to a location in the no wall than the wall condition. Therefore, the wall condition did not exhibit the intended effect of facilitation, but hindered performance. It is possible that the physical barrier of the walls reduced the visibility of the objects. A second possibility is that the walls emphasised categories, at the expense of attention to the relationship between objects. Despite this differentiation at training, TD accuracy at test was similar in the wall and no wall conditions. The differentiation did, however, persist into the test phase for the WS group; level of accuracy was similar to TD 4- to 5-year-olds on the no wall condition, but weaker on the wall condition. As at training, this might be accounted for by more cognitive emphasis on categories, and thus courser accuracy. One could also argue that, as object-location pairing ability was not quite at ceiling for the WS group, this contributed to precision uncertainty at test. However, the same was true of the 4-year-old group, yet performance on the wall condition at test was not disadvantaged. As such, it appears that impairment in the wall, relative to the no wall condition, is unique to WS.

The TD group were not affected by boundary salience. Plumert and Hund (2001) demonstrated superior performance as boundary salience increased. However, the younger participants did not show evidence of dividing the space into quadrants, which could suggest a weaker effect of boundary salience. Given that the TD group here were of comparable or lower CA to Plumert & Hund’s (2001) participants, taken together, it is possible that an effect of boundary salience increases with CA. In contrast, the pattern of superior no wall compared to wall performance in WS is not observed at any point along the typical trajectory in the present experiment or in Plumert and Hund (2001). Thus, the addition of concrete category boundaries did not facilitate the grouping together of locations into categories, but had had a detrimental effect on location memory in WS. This relates to the nature of the deficit in drawing and construction tasks observed in WS. The solutions offered by individuals with WS on such tasks show poor global cohesion (e.g. Bellugi, Sabo & Vaid, 1988). If the spatial layout of the four quadrants is difficult to see as a global whole, as in the wall condition, while the TD children might have overcome this using mental representation, it could be additionally detrimental to an already impaired ability to encode the relations between object locations (i.e. as cohesive sets or categories) in WS.

The TD children overestimated between and within distance estimates. For the wall condition, overestimates were higher for within than between distances. Within distance overestimates became weaker with CA, while between distance estimates remained constant. Clearly, a subdivision effect is not evident in the TD group, however the results are in sequence with the pattern observed by Plumert and Hund (2001), who employed older age groups; they showed consistently overestimated between quadrant distances, while within quadrant distances were overestimated at 7 and 9 years, progressing to no within distance bias at 11 years and in adults.

In the no wall condition, for the TD group, within and between quadrant distances did not differ. We know from the training trials, that this condition was less demanding. Thus it is possible, that progression along the developmental trajectory is more advanced for this condition, hence why the 4- to 8-year-olds here showed a pattern of performance similar to the 7- and 9-year-olds in Plumert and Hund (2001).

The pattern of WS performance showed evidence of a subdivision effect. Between distances were overestimated, but no bias was observed for within distances. The difference between within and between distance estimates was not significant, which might reflect a lack of power, or an attenuated effect. This pattern showed some resemblance to that observed in Plumert and Hund (2001) in their 11-year-old and adult groups. Thus, it appears that although *level* of performance is similar to an early point in typical development (4-year-old children), the *pattern* of location memory performance best matches a much later point in development (11 years onwards). Location memory in WS, therefore, shows deviance rather than delay.

Displacement scores were observed away from the centre of quadrants in the no wall condition only, for both TD and WS groups. This is surprising as this is the less demanding condition and the Category Adjustment model (Huttenlocher et al., 1991) predicts decreased strategy use with increased certainty. However, it is possible that this reflects different strategies at play for each condition, rather than a lack of strategy in the wall condition. A candidate strategy for the TD group in the wall condition is that they were normalising the distances between the objects pairs in each quadrant. Schutte and Spencer (2002) report that young children bias location memory towards an average of those locations. In the present experiment, the objects in each quadrant were two members of a target triad (separated by 6 inches) or one member of a triad and a non-target object (separated by distances of more than 6 inches). Normalising the distances between pairs of objects would have the effect of expanding and reducing the target and non-target within quadrant distances respectively. As this involves pairs of objects, such displacement would not be apparent from the current analysis, as they would cancel each other out. This can also explain the effect of distance estimates observed in the wall condition, as the relative expansion and contraction would cancel out any changes to between quadrant distances. Thus, the effect of normalisation would result in larger within quadrant distances compared to between quadrant distances. As normalisation is a characteristic of young children, development or reduced task complexity dictated a change in strategy. The pattern of performance in the TD group in the less demanding, no wall condition supports this; consistent with Plumert & Hund (2001), the category boundaries (mentally imposed) appear to have been employed as referents resulting in displacement away from each quadrant centre. However, it is difficult to reconcile these displacements with the pattern of similarly overestimated between and within distance estimates for this condition. One can tentatively suggest that displacement might be perpendicular to the distance (between or within quadrant) being measured, resulting in biases that effect distances similarly.

The WS group also showed displacement away from quadrant centres in the no wall condition only. Given that the WS and TD group show different patterns of between and within quadrant distance estimates, this similarity in displacements is puzzling. Plumert and Hund (2001) describe such displacements away from quadrant centres as reflecting a displacement towards the model corners, which then explains the overestimation in between quadrant distances. It is possible that this occurred here. However, first this is contradictory to the pattern of between and within quadrant estimated observed in the TD group in the no wall condition. Second, based on the pattern of between and within distance estimates for the WS group, one would predict similar displacement away from the quadrant centres across both the no wall and wall conditions, yet this was not the case.

Farran and Jarrold (2005) report that the atypical pattern of biases in spatial relations tasks, which are perception tasks, might explain deficits in construction tasks that involve location memory. The current experiment has construction elements as individuals were asked to place objects in the correct locations. In this study, level of performance of the WS group most closely resembled that of a typical 4-year-old. This is younger than the level reached in the spatial relations tasks, which did not differ from matched controls of mean age 6;3 years (Farran & Jarrold, 2005). Thus, the current experiment shows some support for the prediction that the unusual bias observed at perception is more detrimental to construction performance. Thus it appears that poor location memory is a contributing factor to the poor performance observed on production tasks, such as block construction and drawing.

In conclusion, individuals with WS remember locations in an atypical manner; level of performance is particularly weak, even within the poor visuo-spatial domain, yet their pattern of performance has some hallmarks consistent with a subdivision effect. This is surprising, given that in typical development the ability to use subdivision strategies is still developing into late childhood (11 years-old onwards: Plumert & Hund, 2001). In contrast, the TD group in this experiment use a normalisation strategy. The results of this experiment suggest that poor location coding in WS is a contributing factor to their characteristically poor level of performance on construction and drawing tasks.

References

Atkinson, J., Braddick, O., Rose, F. E., Searcy, Y. M., Wattam-Bell, J., & Bellugi, U. (2006). Dorsal-stream motion processing deficits persist into adulthood in Williams syndrome. *Neuropsychologia, 44*, 828-833.

Atkinson, J., King, J., Braddick, O., Nokes, L., Anker, S., & Braddick, F. (1997). A specific deficit of dorsal stream function in Williams syndrome. *Neuroreport: cognitive neuroscience and neuropsychology, 8*, 1919-1922.

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

Farran, E. K., & Jarrold, C. (2003). Visuo-spatial cognition in Williams syndrome: Reviewing and accounting for 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. (2005). Evidence for Unusual Spatial Location Coding in Williams Syndrome: An Explanation for the Local Bias in Visuo-Spatial Construction Tasks? *Brain and Cognition, 59,* 159-172.

Hoffman, J. E., Landau, B., & Pagani, B. (2003). Spatial breakdown in spatial construction: Evidence from eye fixations in children with Williams syndrome. *Cognitive Psychology, 46,* 260-301.

Huttenlocher, J., Hedges, L. V., & Duncan, S. (1991). Categories and Particulars: Prototype effects in estimating spatial location. *Psychological Review, 98,* 352-276.

Jarrold, C., Baddeley, A. D., & Hewes, A. K. (1998). Verbal and Nonverbal Abilities in the Williams Syndrome Phenotype: Evidence for Diverging Development Trajectories. *Journal of Child Psychology and Psychiatry, 39*, 511-523.

Jordan, H., Reiss, J., Hoffman, J. E., & Landau, B. (2002). Intact perception of biological motion in the face of profound spatial deficits: Williams syndrome. *Psychological Science, 13*, 162-167.

Karmiloff-Smith, A. (1997). Crucial Differences Between Developmental Cognitive Neuroscience and Adult Neuropsychology. *Developmental Neuropsychology, 13,* 513-524.

Karmiloff-Smith, A., Grant, J., Berthoud, I., Davies, M., Howlin, P., & Udwin, O. (1997). Language and Williams Syndrome: How Intact is "Intact"? *Child Development, 68*, 274-290.

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

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

Lenhoff, H. M., Wang, P. P., Greenberg, F., & Bellugi, U. (1997). Williams Syndrome and the Brain. *Scientific American*, *277*(6), 42-47.

Mervis. (1999). The Williams Syndrome Cognitive Profile: Strengths, Weaknesses, and Interrelations among Auditory Short Term Memory, Language and Visuospatial Constructive Cognition. In R. Fivush, W. Hirst & E. Winigrad (Eds.), *Essays in honor of Ulric Neisser*. Mahwah, NJ: Erlbaum.

Meyer-Lindenberg, A., Kohn, P., Mervis, C. B., Kippenham, J. S., Olsen, R. A., Morris, C. A., et al. (2004). Neural basis of genetically determined visuospatial construction deficit in Williams syndrome. *Neuron, 43*, 623-631.

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.

Paul, B. M., Stiles, J., Passarotii, A., Bavar, N., & Bellugi, U. (2002). Face and place processing in Williams syndrome: evidence for a dorsal-ventral dissociation. *Neuroreport*, *13*, 1115-1119.

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

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

Schutte, A. R., & Spencer, J. P. (2002). Generalizing the dynamic field theory of the A-not-B error beyond infancy: Three-year-olds' delay- and experience-dependent location memory biases. *Child Development, 73,* 377-404.

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

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

Vicari, S., Bates, E., Caselli, M. C., Pasqaletti, P., Gagliardi, C., Tonucci, F., et al. (2004). Neuropsychological profile of Italians with Williams syndrome: An example of a dissociation between language and cognition. *Journal of the International Neuropsychological Society, 10,* 862-876.

Vicari, S., Bellucci, S., Santa Marinella, G., & Carlesimo, G. A. (2003). Visual and spatial working memory dissociation: evidence from Williams syndrome. *Developmental Medicine and Child Neurology, 45*, 269-273.

Vicari, S., Bellucci, S., & Carlesimo, G.A. (2006). Evidence from two genetic syndromes for the independence of spatial and visual working memory *Developmental Medicine and Child Neurology*

Vicari, S., Brizzolara, D., Carlesimo, G. A., Pezzini, G., & Volterra, V. (1996). Memory in Williams Syndrome. *Cortex*, 32, 502-514.

Viera, A. J., & Garrett, J. M. (2005). Understanding Interobserver Agreement: The Kappa Statistic. *Family Medicine, 37,* 360-363.

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.

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Table 1: Participant CA and RCPM scores, and results of independent t-tests between the WS group and each TD group, on RCPM score.

Group	CA (years; months): mean(SD)	RCPM score: mean(SD)	RCPM, group comparison
WS	23;0 (9;7)	19.80(5.91)	
TD: 4-year-olds	4;2 (0;1)	12.78(4.27)	p=.01 (WS>TD)
TD: 5-year-olds	5;1(0;2)	15.11 (3.10)	p=.03 (WS>TD)
TD: 6-year-olds	6;1(0;3)	16.78(4.76)	p=.13(WS=TD)
TD: 7-year-olds	6;10(0;3)	23.70(3.16)	p=.07 (WS=TD)
TD: 8-year-olds	8;1(0;2)	24.60(3.84)	p=.03 (WS<TD)

Table 2

Correct location placements for training and test phases (max = 8)

Group	Training phase: mean (s.d.)						Test phase: mean (s.d.)	
	Trial 1		Trial 2		Trial 3			
	No wall	Wall	No wall	Wall	No wall	Wall	No wall	Wall
WS	5.00(1.81)	3.67(2.61)	5.50(2.74)	3.67(2.06)	6.25(1.82)	5.17(2.51)	7.45(0.67)	7.27(0.90)
TD: 4-year-olds	5.30(3.20)	3.60(2.27)	6.00(2.36)	3.80(1.99)	5.90(2.56)	5.30(3.23)	6.50(0.97)	6.60(1.07)
TD: 5-year-olds	6.90(1.66)	5.10(1.97))	7.20(1.93)	6.10(2.13)	7.40(0.97)	6.30(2.11)	7.80(0.42)	6.50(2.71)
TD: 6-year-olds	7.50(1.08)	5.90(1.85)	7.30(1.49)	6.60(1.71)	7.20(1.93)	7.30(1.15)	7.80(0.42)	7.70(1.84)
TD: 7-year-olds	7.50(1.58)	6.90(1.52)	8.00(0.00)	6.9(2.02)	8.00(0.00)	7.20(1.39)	7.90(0.32)	8.00(0.00)
TD: 8-year-olds	8.00(0.00)	8.00(0.00)	8.00(0.00)	8.00(0.00)	8.00(0.00)	8.00(0.00)	8.00(0.00)	8.00(0.00)

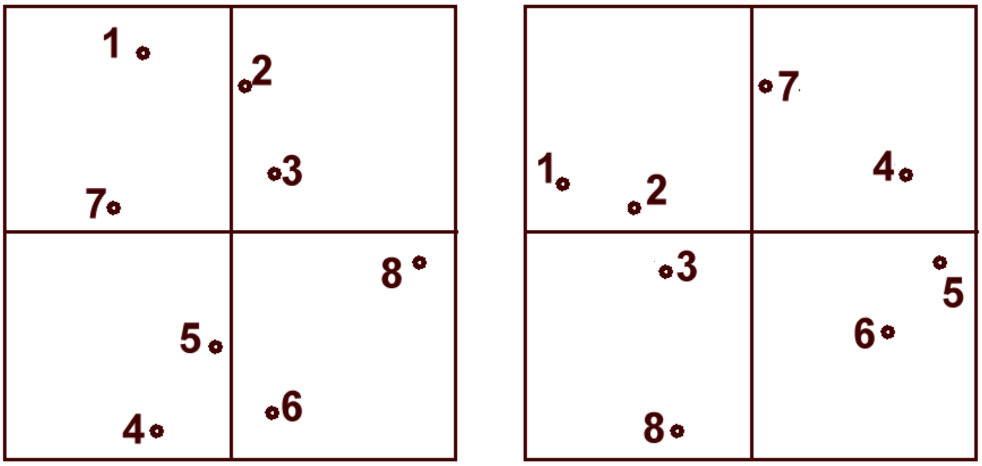
Figure captions

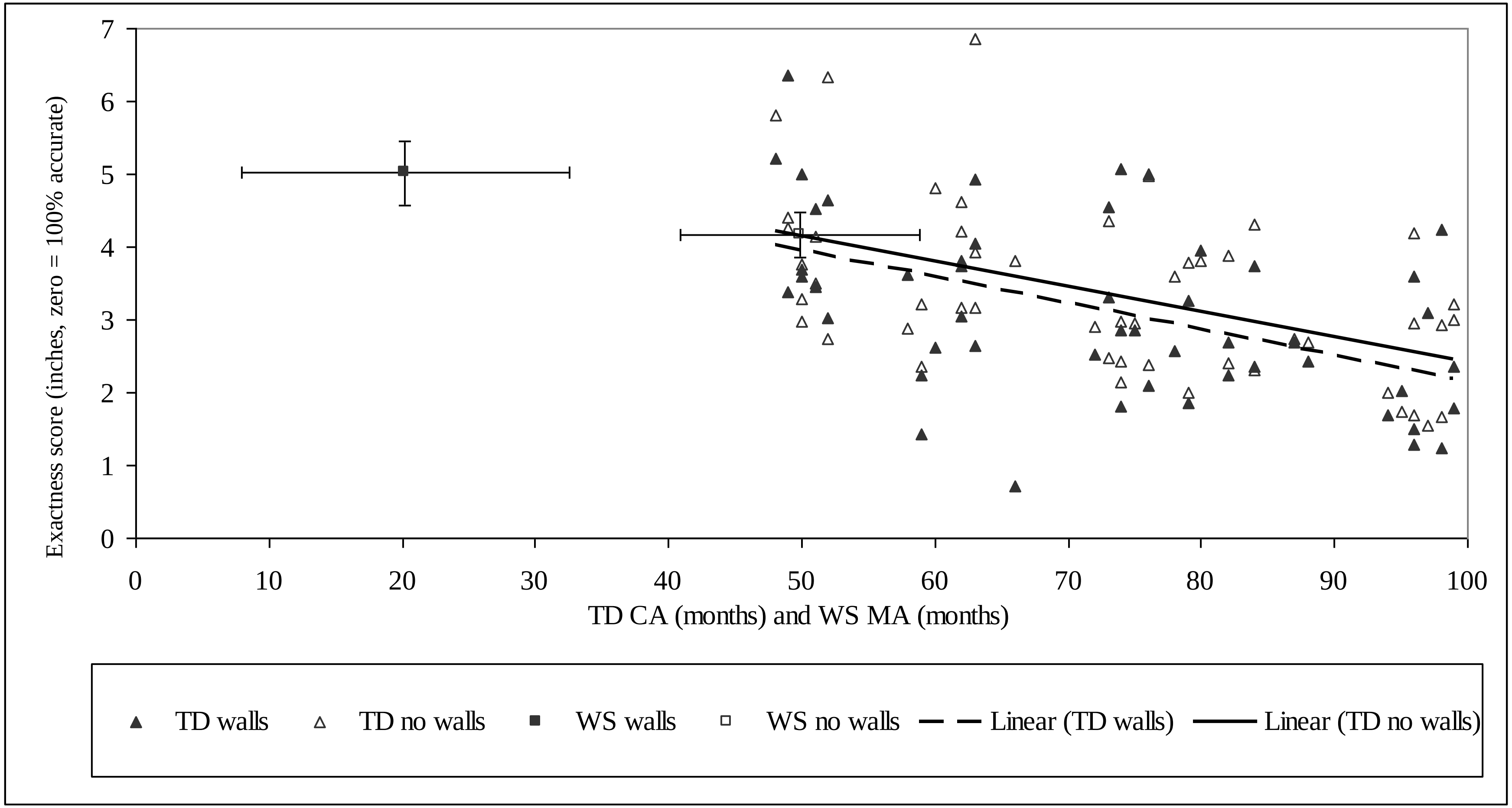
Figures 1a and 1b: Target locations

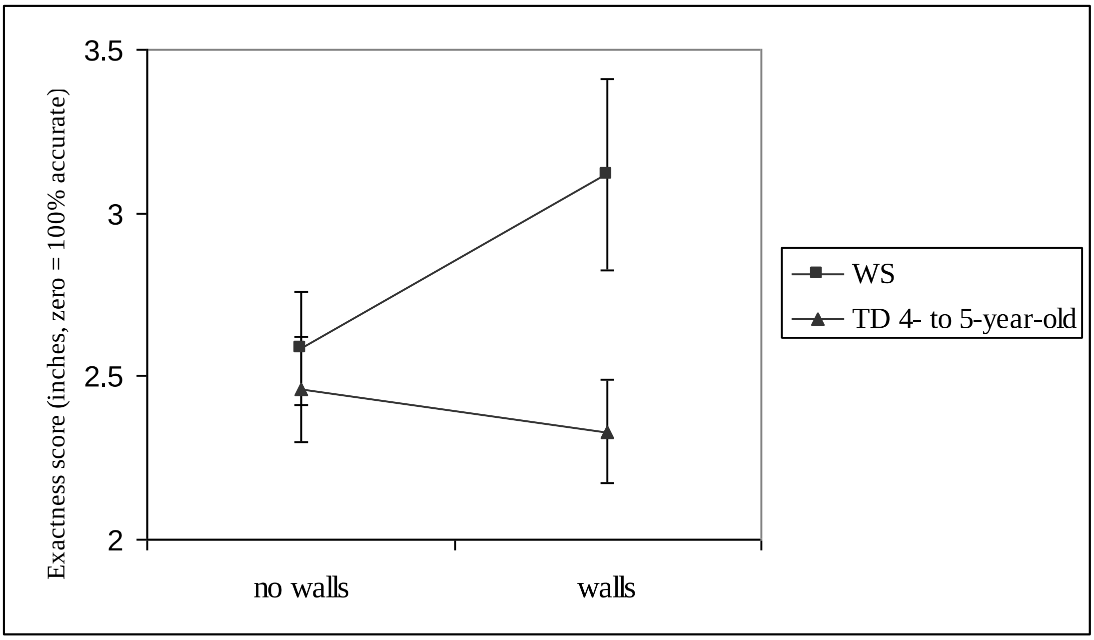
Figure 2: Exactness scores plotted against age: TD individual participant scores against Chronological age (CA); WS mean scores with X and Y standard error bars, against Mental age (MA; predicted by the TD trajectory)

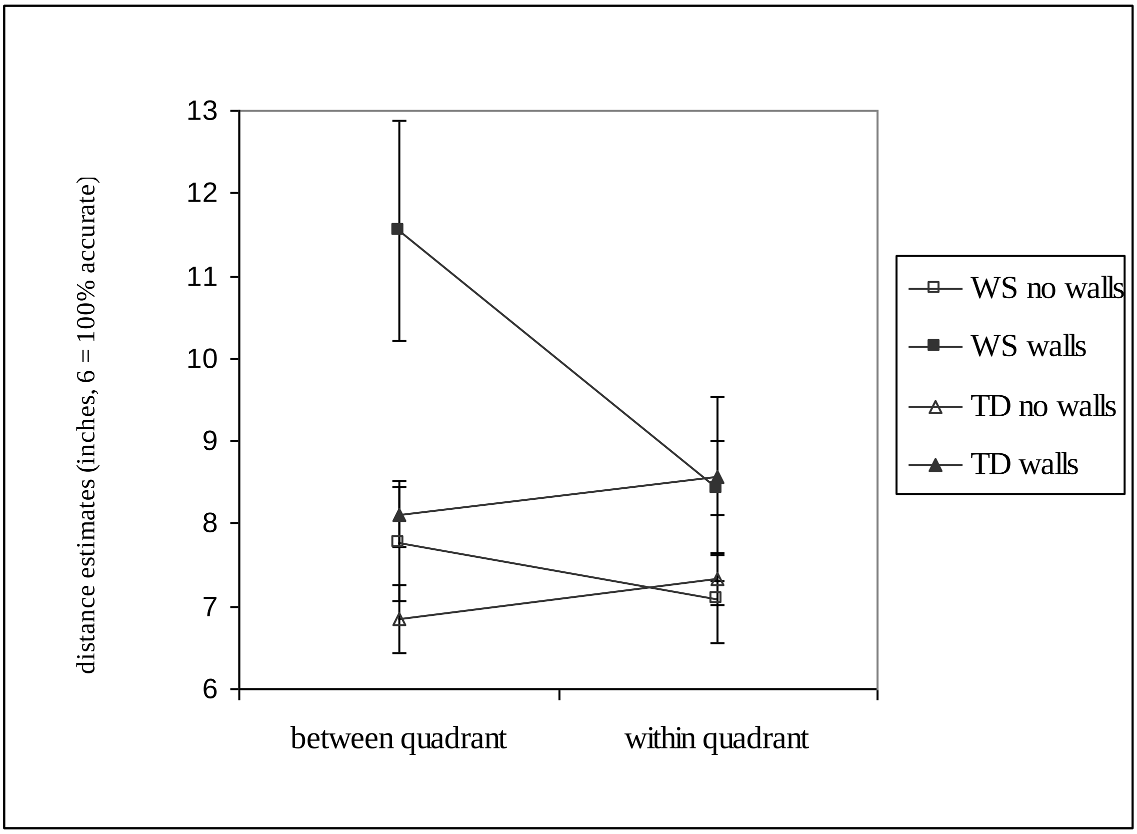
Figure 3: Mean exactness scores

Figure 4: Mean distance estimates


1
2
3
7
8
5
6
4
7
1
4
2
3
5
6
8


Exactness score (inches, zero = 100% accurate)
7
6
5
4
3
2
1
0
0
10
20
30
40
50
60
70
80
90
100
TD CA (months) and WS MA (months)
TD walls
TD no walls
WS walls
WS no walls
Linear (TD walls)
Linear (TD no walls)


3.5
3
2.5
2
Exactness score (inches, zero = 100% accurate)
no walls
walls
WS
TD 4- to 5-year-old


distance estimates (inches, 6 = 100% accurate)
13
12
11
10
9
8
7
6
between quadrant
within quadrant
WS no walls
WS walls
TD no walls
TD walls