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Visuo-Spatial Cognition in Williams Syndrome: Reviewing and Accounting for the Strengths and Weaknesses in Performance

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Abstract

Individuals with Williams syndrome typically show relatively poor visuo-spatial abilities in comparison to stronger verbal skills. However, individuals' level of performance is not consistent across all visuo-spatial tasks. The studies assessing visuo-spatial functioning in Williams syndrome are critically reviewed, in order to provide a clear pattern of the relative difficulty of these tasks. This prompts a possible explanation of the variability in performance seen which focuses on the processing demands of some of these tasks. Individuals with Williams syndrome show an atypical processing style on tests of construction, which does not affect tests of perception.

Introduction

Ever since the pioneering work of Bellugi and her colleagues (e.g., Bellugi, Sabo, & Vaid, 1988) it has been clear that a fundamental aspect of the psychological profile of individuals with Williams syndrome (WS) is their relatively poor performance on tests of visuo-spatial cognition (see Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995; Mervis, 1999). Many researchers describe the cognitive profile of WS by detailing the marked contrast that is seen between these individuals' verbal and visuo-spatial abilities. For example, Bellugi, Wang, and Jernigan (1994) describe a "Pattern of linguistic preservation and marked spatial cognitive deficit" (p. 44), whilst Udwin and Yule (1991) suggest that "overall their verbal abilities are markedly superior to their visuo-spatial and motor skills" (p. 233). In general it is the case that the verbal abilities of individuals with WS are superior to their non-verbal abilities (Grant et al., 1997; Howlin, Davies & Udwin, 1998). However, Jarrold, Baddeley, & Hewes (1998) and Jarrold, Baddeley, Hewes, & Phillips (in press) claim from cross-sectional and longitudinal data respectively, that in WS, verbal ability improves at a faster rate than non-verbal ability, so that as individuals develop an increasing discrepancy between these two domains emerges. This is supported by Atkinson et al. (in press) who report steeper slopes in improvement with age in vocabulary and grammar ability, than the comparatively slow rate of improvement in ability with age on three visuo-spatial tasks. While the discrepancy between verbal and non-verbal ability is, broadly speaking, characteristic of WS, the situation is complicated by the considerable variance between the composite measures of any IQ score. These composite scores often hide an interesting pattern of differences in abilities shown on tests that measure particular aspects of cognition within the verbal or visuo-spatial domains. Karmiloff-Smith et al. (1997) have shown, for example, that performance is not uniform on a range of tasks measuring different aspects of verbal ability. The purpose of this article is to critically review the research carried out to date in the area of visuo-spatial cognition in WS, with the aim of explaining the reasons for the varying levels of performance between different visuo-spatial tasks. As this is arguably the weakest of all aspects of cognition in WS it is particularly important to provide potential explanations for the difficulties these individuals encounter on visual and spatial tests.

Methodological Issues

Methodological issues need to be taken into account when evaluating the findings of any study; however, particular methodological problems arise when working with special populations in general, and with individuals with WS specifically. Consequently, before reviewing the studies of visuo-spatial cognition in WS we provide a brief outline of these methodological concerns and their potential effects on these studies.

Floor and ceiling effects can be a major problem when testing atypical populations. Clearly when either effect occurs, the test used may be artificially constraining the possible range of performance. Floor effects are particularly prevalent when testing visuo-spatial processing in WS, as this is such a weak area of cognition. This is evident in studies employing the Benton Lines Orientation test to assess individuals with WS (Benton, Varney, & Hamsher, 1978; see Bellugi et al., 1988; Rossen, Klima, Bellugi, Bihrle, & Jones, 1996; Wang, Doherty, Rourke, & Bellugi, 1995). Such results can only tell us that a group's abilities are at or below the lowest level that the test purports to measure, and thus that the group's scores may not be truly representative of their actual skills. In the present context, ceiling effects are commonly seen in comparison or control groups. When WS participants are matched to a control group for chronological age (CA), it is difficult to find a test which encompasses the range of abilities seen across the two groups. If the test is too easy for the control group they will score at ceiling, and as a consequence the performance of the WS group may erroneously appear to be close to that of controls. This possibility will be discussed in relation to much of the research that addresses face recognition in WS, where CA matched control groups are often employed (e.g. Karmiloff-Smith, 1997).

The problem of floor and ceiling effects can be overcome somewhat by matching individuals with WS to typically developing (TD) groups for mental age (MA). This approach has been adopted by Bertrand, Mervis, and Eisenberg (1997) for example who, in addition to typically developing CA matched controls, also employed a typically developing control group matched for MA. This reduces the problem of differing levels of ability, but creates a new concern due to the discrepancy that will necessarily arise between the CAs of each group. The higher CA of the WS group will equate to them also having more experience, more practice in using their skills and more strategic coping styles (although this may also be linked to MA) which can introduce confounds into the experiment. This can be overcome by employing groups of individuals with learning difficulties as controls (Crisco, Dobbs, & Mulhern, 1988), or in addition to TD controls (Jarrold, Baddeley, & Hewes, 1999), because these individuals can potentially be matched to WS groups for both CA and MA.

However, an additional issue that arises whenever a control group is employed concerns the criteria used to match groups. This follows from the uneven profile of abilities of individuals with WS in contrast to the flat profile seen in typical development. A CA matched control group is likely to differ from a WS group in all areas of intelligence, but more so in the visuo-spatial domain and less so in the verbal domain. When matching groups by general MA, a control group will have higher visuo-spatial skills, and lower verbal skills than the WS group. Any discrepancies in results that then emerges between groups could be primarily due to these differences in levels of ability, rather than in performance on the task in question. Visuo-spatial cognition in WS is particularly susceptible to the problems of matching by general MA, as level of ability in this area is so low.

A related problem occurs when test batteries such as the Wechsler scales (Wechsler, 1974, 1981) or the Differential Ability Scales (DAS; Elliot, 1990) are employed. These batteries use a large number of tests to determine an individual's full scale IQ (FSIQ) or level of development (MA). The uneven profile of abilities in WS means that performance will not be equivalent across all of the individual subtests of a test battery. It is therefore important to consider the number and type of subtests used in order to determine which specific abilities are contributing to the composite measure of FSIQ. The Wechsler scales contain 5 non-verbal and 5 verbal subtests, whilst the DAS contains 6 subtests in total. Shortened versions of the WISC are also used (e.g., Grant et al. 1997). Speculatively, the more subtests employed, the more likely the average score will encompass the full range of abilities, thus producing a more reliable measure of general ability. However, although FSIQ can reliably assess general ability, due to the imbalance in skill in individuals with WS, an IQ score is not necessarily a particularly valid measure. It merely represents an averaged score of many differing levels of ability, and is therefore unlikely to be a powerful predictor of functioning on other tests of interest (as in typical development). In the case of WS, the individual subtests arguably provide more informative details of cognition than a composite IQ score.

These problems of matching are exacerbated when individuals with learning difficulties are used as controls, because the cognitive profile of these controls may also be less uniform than that seen in typical development. For example, individuals with Down syndrome (DS) are often used as controls for individuals with WS (e.g., Bellugi, Bihrle, Neville, Doherty, & Jernigan, 1992; Bellugi et al., 1988; Rossen et al., 1996; Wang et al., 1995). The choice of DS controls is based on the assumption that these individuals exhibit a flat profile of abilities. However, Klein and Mervis (1999) present evidence against this assumption. They suggest that DS individuals have a relative strength in the area of visuo-spatial construction, and a relative weakness in verbal ability (see also Chapman, 1995; Fowler, 1990; Jarrold & Baddeley, 1997; Miller, 1987). In light of this, where studies have used DS individuals as controls, any observed differences in scores could be due to strengths or weaknesses in the DS control group as much as in the WS group.

To overcome these difficulties control groups can be matched by their performance on a single measure. In order for matching to be appropriate the measure used must be drawn from the same area of cognition as the area under investigation. This ensures that the scores of the control group are predictive of the expected level of performance of the WS group in the test condition, although none of the studies reviewed here adopt this approach. A further problem can arise if the predictive MA measure and the testing measure are too closely related. In this case the experimenter might just be testing the same abilities twice in both groups, and any interesting results that might indicate a deviation from typical development will be wiped out (Bishop, 1997). Bishop claims that even when a difference is noted in these cases, one cannot be sure that this is not simply due to differences in the relative reliability of the two tasks.

In summary, every method has some weaknesses, and research with individuals with WS is particularly susceptible to the problems of floor and ceiling effects and of matching controls appropriately. This emphasises the importance of employing a number of methodological techniques in a single study, with the intention of counteracting the weaknesses of one methodology with the strengths of another. For example, matching by both MA and by CA as in Bertrand et al. (1997), or by employing both TD controls and controls with moderate learning difficulties (see Jarrold et al., 1999). In the following two sections of the paper we review the main body of research in the area of visuo-spatial cognition in WS, discussing first studies which have used test batteries such as the Wechsler scales, and secondly those employing tests of specific aspects of visuo-spatial ability. Clearly all of these studies need to be interpreted in the light of the methodological concerns raised here.

Studies Employing Standardised Test Batteries

Non-verbal Subtests of the WAIS / WISC

The WS cognitive profile has been primarily documented using standardised test batteries such as the Wechsler Intelligence Scale for Children and the Wechsler Adult Intelligence Scale (WISC-R, WAIS-R; Wechsler, 1974, 1981). Five studies provide information on the individual scores of each subtest of the battery (Arnold et al., 1985; Dall'oglio & Milani, 1995; Howlin et al., 1998; Udwin & Yule, 1991; Udwin, Yule & Martin, 1987), although the sample employed by Udwin and Yule (1991) is a subset of that employed by Udwin et al. (1987). Mean subtest scores where provided (2 studies: Howlin et al., 1998; Udwin et al., 1987) are given in Table 1. The five non-verbal subtests of the WISC-R and the WAIS-R provide a measure of Performance IQ (PIQ). These are: Picture Completion; where a picture is presented and the participant has to indicate what is missing; Picture Arrangement, which involves placing a series of pictures into a sequential order of events; Block Design, where the participant is instructed to use coloured blocks to model an example pattern; Object Assembly, which is a jigsaw type task; and Coding which is a timed task where the participant uses a key to draw specified symbols below a set of numbers.

Dall'oglio and Milani (1995) assessed 16 individuals with WS aged 4;10 to 15;4 years (no mean age given) using the WISC-R. Their participants showed poor performance on the Block Design, Coding, and Picture Arrangement subtests, in contrast to better performance on the Picture Completion and Object Assembly subtests. Arnold et al. (1985) measured the performance of 23 participants (mean age: 10;4 years, range: 7;2 to 13;1). They report that Coding was significantly poorer than Picture Completion, Block Design, and Object

Assembly. In addition, performance on Picture Completion was higher than Picture Arrangement performance, a result which is consistent with the data presented by Dall'oglio and Milani (1995).

Howlin et al. (1998) studied 62 individuals with WS who were of mean age: 26.5 years (range: 19 to 39 years). In contrast to the studies described above, they found that scores on the Picture Arrangement task were the highest amongst the PIQ subtests and was significantly higher than scores on the Coding subtest (referred to as Digit Symbol) where the lowest scores were achieved (see Table 1).

Udwin et al. (1987) originally tested 44 participants with a mean age of 11;1 years (range 6;0 to 15;9 years). They did not find highest scores on the Picture Arrangement task, but a subsequent study of 20 of these individuals, who had a mean age of 10;4 years (range: 6;5 to 14;5 years) did achieve their highest mean score on the Picture Arrangement subtest (Udwin & Yule, 1991). In both studies individuals showed significantly poorer ability in Coding than a combined mean of the scores achieved on the other four Performance subtests (see Table 1).

Table 1 about here

Although the profile of scores on the five non-verbal subtests of the WISC and WAIS is not entirely consistent across these studies, it is clear that scores on the Block Design and Coding subtests tend to be among the lowest obtained, while the Picture Completion and Object Assembly tasks produce consistently higher scores. The ranked position of the Picture Arrangement task is less consistent, with performance varying from a central to a higher position in comparison to other subtests.

A related study (Atkinson et al., in press) employed 3 tasks, two of which were an Object Assembly subtest (taken from the Wechsler Pre-school and Primary Scale of Intelligence-Revised (WPPSI-R; Wechsler, 1989)), and a block construction task (Atkinson, Macpherson, Rae, & Hues, 1994). The performance of 73 children with WS (mean age: 7;3 years; range 8 months to 13;7years), was compared against a set of norms obtained in a previous study (Atkinson et al., 1994). Graphical information clearly indicates that the performance of individuals was poor on these tasks with respect to age norms. Thus, the performance of the large sample employed in this study concurs with those of the studies reported above.

Non-verbal Subtests of the DAS

The DAS consists of two alternative batteries: a Pre-school battery (ages 3;6 to 6;11) and a School-age battery (ages 5;0 to 17;11). The Pre-school version has six subtests, three of which are used to derive non-verbal mental age. These are; Picture Similarities, which involves the participant matching a sample card to one of four pictures based on perceptual similarity or semantic association; Pattern Construction, a task similar to the Block Design subtest of the Wechsler scales; and Copying where the individual copies line drawings. Four School-age measures, two labelled as non-verbal, and two labelled as spatial, are employed to ascertain non-verbal ability. The non-verbal tasks are Matrices, where the correct design has to be selected to complete a matrix pattern; and Sequential and Quantitative Reasoning, in which items such as a shape have to be selected to complete a sequence of items. The spatial tasks are Pattern Construction as above, and Recall of Designs in which the individual is shown an abstract design for a period of 5 seconds, after which it is removed, and the child is requested to draw it from memory.

Jarrold et al. (1998) compared selected subtest scores of the DAS battery in their sample of 16 individuals with WS (mean age 16;9 years, range 6;11 to 28;0). Among those individuals functioning at the Pre-School level of the DAS the level of performance on the non-verbal tasks employed, ranked in descending order, were as follows: Copying, Picture Similarities, and Pattern Construction. The corresponding order, on the 3 subtests employed, for individuals at the School-Age level was: Matrices, Sequential and Quantitative Reasoning, and Pattern Construction. The relatively low performance on the Pattern Construction subtest in both DAS batteries is consistent with the results of Block Design performance in the Wechsler studies described above.

Summary: The WS Cognitive Profile

The uniformly poor performance seen on the Pattern Construction and Block Design tasks in the DAS and Wechsler test batteries suggest that the skills needed for this kind of task are particularly weak in WS. This has lead to the test being investigated as a single measure rather than as part of a composite of subtest scores. Mervis, Morris, Bertrand, and Robinson (1999) assessed performance on the Pattern Construction subtest of the DAS in 80 WS participants (age range: 4 to 47 years). 80% of the sample had a score which corresponded to the 1st percentile or lower for typical performance. 58% of participants were at floor on the task, which may be masking even poorer abilities. Only 10% scored within the normal range. Other studies (e.g. Frangiskakis et al., 1996; Bellugi et al., 1988, 1992) have also reported similarly low scores on block construction tasks.

Mervis (1999) has used the consistent weakness in Pattern Construction, along with other characteristics of WS, as a basis for a set of psychological criteria for diagnosing WS. These are a "definite strength in auditory short term memory, relative strength in language, and extreme weakness in visuo-spatial construction" (Mervis, 1999, p.197). The Pattern Construction test is used as the measure of visuo-spatial construction ability and is involved in two of the criteria. Performance on this test must be below the general MA measure; and also below Digit recall score (a measure of verbal short term memory). A number of studies have found high levels of sensitivity and specificity using these criteria to identify individuals who do and do not have WS (Frangiskakis et al., 1996; Jarrold et al., 1998; Mervis, 1999; Mervis et al., 1999; Mervis et al., 2000).

Studies Employing Specific Visuo-Spatial Tests

Tests of Spatial Organisation

Bellugi and colleagues (e.g., Bellugi et al., 1988, 1992, 1994) have suggested that individuals with WS process information at the local level, i.e. they focus on the parts of an image rather than its whole. The basis for this claim is the pattern of errors shown by some individuals with WS on the Block Design task. Rather than recreating the overall spatial organisation of the pattern, i.e. four blocks in a 2 by 2 square arrangement, individuals with WS may select the appropriate individual blocks, but place them in an unorganised manner which fails to maintain the relationships between blocks. There are a number of factors to take into account when considering this suggestion of a local processing bias. Firstly, young children also produce solutions where the configuration are broken (e.g., Akshoomoff &

Stiles, 1996) which suggests that the errors made by individuals with WS might not be due to a deviant processing style. Secondly, other authors have suggested that a local processing approach should actually be beneficial to performance on the Block Design test (Happé, 1994, 1999; Shah & Frith, 1993). This is because to succeed on this task the individual needs to resist the gestalt form of the overall pattern, and analyse the stimulus in terms of its component parts. In support of this, Shah and Frith (1993) have shown that typically developing individuals complete test items more rapidly when the stimulus is presegmented into its constituent blocks rather than presented as a whole. In addition, individuals with autism, who are known to show a local visual processing bias on many tasks (Happé, 1999), show particularly strong levels of performance on the Block Design test precisely because the task benefits from a local analysis (Happé, 1994; Shah & Frith, 1993).

If individuals with WS do process information at a relatively local level, then this form of segmentation should not cause response time to decrease. Mervis et al. (1999) investigated the performance of a group of 21 individuals with WS of mean age 29.5 years, using both a standard and a segmented version of the DAS Block Design task. Response times were generally reduced when blocks were segmented. This facilitation suggests that individuals with WS do not rely exclusively on local processing in the standard Block Design task as advocated by Bellugi. Consequently, the authors put forward the suggestion that: "individuals with Williams syndrome have difficulty segmenting the whole into its component parts" (p.94).

Farran, Jarrold, and Gathercole (2001) were interested in the magnitude of this facilitation effect, i.e. whether the effect of segmentation observed in WS on this type of task is to a greater, equal or lesser extent than the typical population. The authors employed a novel task, the Squares task, which is a 2 dimensional version of the Block Design task. Individuals were presented with either a segmented or a non-segmented version of a model image, which could be copied by placing four squares in the correct 2 by 2 formation. Each square was divided centrally into two colours either across the diagonal (oblique squares) or vertically/ horizontally (non-oblique squares). The performance of 21 individuals with WS of mean age: 19;11 years (range 9;6 to 38;5 months) was compared to that of 21 TD controls matched individually by score on the Ravens Coloured Progressive Matrices (RCPM; Raven, 1993). Results showed that both groups were equally facilitated by segmentation. This implies, not only that individuals with WS do not show a local processing bias, but that their processing preferences appear to be entirely typical.

Hoffman, Landau, and Pagani (in press) were interested in the process of construction in WS. Hence, they investigated the pattern of eye fixations whilst participants were completing a computerised block construction task. They report that, in complex puzzles, 8 individuals with WS (mean age: 9;5 years, range: 7;0 to 13;11) were just as able to detect an error in their solution as a group of 8 TD controls matched for IQ on the Kaufman Brief intelligence Test (KBIT; Kaufman & Kaufman, 1990) of mean age 5;3 years (range: 5;1 to 6;4). However, the WS group were less accurate at choosing the correct puzzle piece, checked their partial solutions less often, and were less likely to change their solution when they detected an error. The authors suggest that these factors account for the significantly lower accuracy overall in the WS group than the control group. This suggests that it is the process of block construction, rather than the perception of the model image which effects WS performance.

Pani, Mervis, and Robinson (1999) administered a visual search task in order to test whether individuals with WS are influenced by global information. They employed a task, taken from Banks and Prinzmetal (1976), in which the participant is asked to indicate whether a “T” or an “F” is present amongst the stimuli in a visual array. These targets are presented alongside distracters, which were described as halfway between a "T" and an "F". This task affects the efficiency of global and local processing approaches depending on how the stimuli are manipulated. Stimulus grouping particularly affects individuals who adopt a relative global processing approach to the task. In contrast individuals employing a relatively local approach are more likely to be affected by the number of distracters in the array. Banks and Prinzmetal (1976) found that response times (RT) were significantly faster when targets were isolated and the distracters were grouped by proximity than when there were fewer, more evenly spread stimuli. In other words, the effect of gestalt grouping was stronger than the effect of display size, implying a predominance of global over local processing in TD adults. Pani et al presented this task to 12 individuals with WS, with a mean age of 30;11

years (range: 19;3 to 47;6), who were matched to 12 TD individuals, by gender and CA. Both groups were more influenced by gestalt grouping than by display size, which suggests that individuals with WS have a global processing precedence as seen in typical development. The WS group were less influenced by the number of distracters than the controls. The authors argue that this implies that individuals with WS are less able to disengage from global processing than the controls. The suggestion made by Bellugi and colleagues that individuals with WS have a global processing deficit is therefore not supported by this study.

Farran and Jarrold (submitted a) investigated the comprehension of spatial relations in WS. Global accuracy is dependent on reproducing the spatial relations between the parts of the image, therefore spatial relations are particularly important in tasks in which an image must be reproduced. Twenty-one individuals with WS of mean age 21;2 years, and 21 TD controls of mean age 6;3 years were matched individually by level of performance on the RCPM. Three tasks were administered, two tasks measured categorical spatial relations and co-ordinate spatial relations respectively, and a third task measured the comprehension of visual relations. Categorical spatial relations refer to linguistic categories such as “next to”, “to the left of” and “above”, and are used to describe the spatial layout of a scene. Co-ordinate spatial relations refer to distances, and are useful for spatial navigation. In both of the spatial relations tasks, participants were shown an image of a man holding a bat. Following the appearance of a ball, participants were asked whether the ball was ‘above’ or ‘below’ the bat (categorical relations), or whether the ball was ‘in’ or ‘out’ (co-ordinate relations). In the visual relations task, the individual was presented with 3 coloured squares. The two outer squares were blue and green respectively and the middle square varied in hue between blue and green. Participants were asked to judge whether the middle square was more like the green square or more like the blue square. In all three tasks, experimental trials followed a set of 12 practice/ training trials. Results showed that the WS group, although showing a similar pattern of performance to the TD controls, were significantly poorer than the TD controls in completing both the spatial relations and the visual relations tasks. This suggests that individuals with WS find it difficult to comprehend both the spatial relationship between elements of an image, and also variations in colour hue, which are also important indicators of spatial organisation. It is possible that this apparent deficit in spatial relational understanding has a negative affect on the ability of individuals with WS to reproduce images accurately.

Tests of Drawing

Bellugi et al. (1988) employed a subtest of the Boston Diagnostic Aphasia Examination (BDAE; Goodglass & Kaplan, 1972) to assess drawing skills in WS. In this task the individual is asked to draw a set of common objects, first from memory and then using a model. The objects become increasingly complex (i.e. cross, cube, flower, house) as the task progresses. Bellugi et al. (1988) observed that 3 individuals with WS, aged 11, 15 and 16 years, drew parts without integrating the drawings into functional objects; drawings also lacked representation of depth and perspective. These authors also administered the Developmental Test of Visual-Motor Integration (VMI; Beery & Buktenica, 1967), which consists of a set of 24 geometric figures of varying complexity which have to be copied. These are divided into four categories, which are, in order of increasing difficulty: single lines, simple shapes, intersection of lines, and items involving integration of 2 or more shapes. The 3 individuals with WS in this study again lacked the ability to organise their drawings. Of the 24 figures, only the first 8, belonging to the single lines and simple shapes categories, were completed by all three children. This is the level that a child aged 4;11 years would be expected to reach and was considerably below these participants' CA. Bellugi et al. (1988) interpret these results as an " inability to maintain two hierarchically organised levels in their drawings" (p.295).

Wang et al. (1995) employed the VMI in a study with 10 participants with WS (mean age: 15.7 years, range: 11 to 18 years and FSIQ: 48.9) and a control group of individuals with DS matched for CA and FSIQ. The WS participants successfully completed a mean of 7.5 figures; a level significantly below that obtained by the DS group, although this may be due to elevated performance among the DS group as their visuo-spatial construction abilities are stronger than their verbal abilities (see Klein & Mervis, 1999). The authors suggest that the individual drawings of the individuals with WS indicate that qualitatively there is "an impairment in global coherence" (p.58) and that " local features were not oriented correctly with respect to each other." (p.59).

Bertrand et al.(1997) also employed the object drawing subtest of the BDAE and a reduced version of the VMI (Beery, 1989). In study one, eighteen children with WS of mean CA: 9;11 years (range 9;2 to 10;7), and mean MA: 5;6 (range: 3;0 to 7;0) participated and were compared to two control groups of TD individuals. One control group was matched by MA (mean CA: 5;6, range 3;5 to 6;11) and the other by CA (mean CA: 9;11 years, range 9;2 to 10;8). The performance level of the WS group on the VMI was equivalent to that of typical children of 4;10 years, and was significantly lower than that observed in both control groups. On the BDAE, individuals with WS produced significantly fewer recognisable drawings, fewer major parts and more disorganised drawings than CA matched controls. Recognisability and disorganisation of WS drawings was not significantly different from that of the MA matched controls' drawings, although significantly fewer major parts were produced by the WS group. In a second study, Bertrand et al. (1997) investigated the developmental progression of typically developing individuals aged 4 to 7 years on the above two tests. Results from the VMI revealed that copying geometric forms that require the integration of component parts is a skill that is not fully acquired until 6 years. The drawings of 4-year-old children and many of the 5-year-olds were unintegrated resembling those of the WS group in study one. The drawings from the BDAE of older children were significantly more recognisable, included more major parts were more organised than those of younger children Again, the level of disorganisation at four years resembled that of the individuals with WS in study 1. Bertrand and Mervis (1996) report developmental improvements in the drawings of six adolescents with WS between two testing points, one at age 9 to10 years and the second at 12 to 14 years, which followed the same path as typically developing individuals.

The Delis Hierarchical processing task, also known as the Navon task (Navon, 1977) is another task which can involve drawing. This test focuses more directly on the issue of global and local levels of processing. A figure is presented which consists of local features such as small L's which when seen as a whole, make up a larger shape or letter such as a D. The individual is invited to draw the figure from memory or to copy it. In each case both local and global processing are required if both the parts and the whole are to be drawn accurately.

Bihrle, Bellugi, Delis, and Marks (1989) compared the performance on this task of 14 individuals with WS, mean age: 13.12 (range: 9 to 18 years), mean IQ: 57.42 (range: 49 to 77), with that of a group of 10 CA matched TD controls and 9 children with DS matched for CA and IQ. In the memory condition, the TD group performed significantly better than both WS and DS children. However, of particular interest is a significant interaction between group and hierarchical level that emerged from the analysis. Individuals with WS were significantly more accurate at drawing local features relative to global figures whilst individuals with DS showed the opposite pattern. WS participants omitted significantly more global forms than both DS and TD groups. In the copy condition the same group by hierarchical level interaction emerged. The TD group were equally competent at copying both global and local forms, although this may be because they performed at ceiling on the test. Because of this it is possible that the significant interactions reported in this study are driven by atypical performance in the DS group, rather than the WS group. Entirely similar results were reported on this task by Rossen et al. (1996) who compared the performance of 6 individuals with WS (mean age 14;2, mean IQ: 50.8) to that of 6 individuals with DS matched for CA and IQ. No statistical analysis is given in this paper and comparison with a TD control group was not made, so once again it is difficult to establish whether the performance of the WS group is atypical. However, these two studies point towards a possible preference for local processing in WS in drawing.

The Delis task was also employed by Stevens (1997), who asked 13 individuals with WS, of mean CA: 18;10, and mean MA: 5;3, to both draw the stimuli themselves and to give verbal instructions to another person as to how to draw each figure. Stevens did not find that the WS group as a whole showed a local bias in their drawings, although this was seen among the drawings of five of the 13 individuals with WS. When asked to give instructions to someone else there was much less evidence of any local bias. In this case only one individual showed a consistent local preference, and their responses contained some global elements. As a result Stevens suggests that any local bias seen on the Delis task is unlikely to be due to an abnormality of visuo-spatial perception, but that it could emerge during the planning or execution of a motor response.

Farran, Jarrold, and Gathercole (submitted) compared the drawing ability of the Navon figures to perception of the same figures in WS. Twenty-one individuals with WS of mean age 20;9 years (range: 10;2 to 39;2) and 21 TD participants of mean age; 6;7 years (range: 5;9 to 7;9) were matched individually by performance on the RCPM. Performance on the drawing task showed significantly better local accuracy than global accuracy in the WS group, whilst the performance of the control group was comparable in local and global accuracy, thus replicating the results of previous studies (Bihrle et al., 1989; Rossen et al.,1996). Two perceptual versions of the Navon task, measuring divided and selective attention respectively, were adapted from Plaisted, Swettenham, and Rees (1999). In the divided attention task, the individual was required to indicate whether the large letter (global level) or the small letter (local level) was a letter A, and hence were required to switch attention across the hierarchical levels. In the selective attention task, attention was focused on one hierarchical level at a time. In one condition, participants were required to focus at the global level and had to indicate whether the large letter was an H or an S. In the other condition, attention was directed to the local level and participants indicated whether the small letter was an S or an H. In contrast to the results from the drawing task, Farran et al. found that at the perceptual level, individuals with WS showed the same pattern of performance as the TD controls, with no evidence of either a global or a local processing preference. This suggests that the local preference seen in the drawing abilities of individuals with WS does not reflect a local perceptual processing bias.

Tests of Visual Closure

Perceptual closure is the ability to use fragmented information to obtain a configural percept. This is essential in cases of object recognition when information about the object's global configuration is incomplete. In these instances the individual needs to construct a global percept from the available information, rather than focusing on the local, fragmented elements of the stimulus. A well known example of a test of visual closure is the Mooney faces test (Mooney, 1957), in which the participant must discriminate between faces and nonfaces that have highly exaggerated shadows and highlights. Bellugi et al.(1988) presented this test to their three individuals with WS and observed higher performance levels than would be expected for these individuals' CA. They took this to indicate " intact abilities to perceive and differentiate shape and form" (p.293). Wang et al. (1995) also gave the Mooney faces test to their 10 individuals with WS. In contrast to Bellugi et al.'s results, they found that performance on this and other tests of visual closure was not significantly different from that of a comparison group of 9 individuals with DS matched for CA and IQ. The performance of both groups fell within the range expected for pre-school and young school-aged children which was consistent with these individuals' mental age levels. Although both of these studies employ relatively small samples, and lack a control group other than individuals with DS, their results suggest that individuals with WS may be able to perceive the global aspects of an image.

Tests of Face Recognition

Face recognition is viewed as an area of relative strength within the domain of visuo-spatial cognition in WS (e.g., Bellugi et al., 1988; Rossen et al., 1996). The Benton Test of Facial Recognition (Benton, Hamsher, Varney, & Spreen, 1983) has been administered in a number of studies (Bellugi et al., 1988, 1992; Karmiloff-Smith, 1997; Rossen et al., 1996; Wang et al., 1995). In this test the participant is initially shown a front-view photograph of a face, and is then asked to identify this target from among distracters. In a subsequent section of the task, the target and distracters are presented in three-quarter view, and in the final section the target appears among faces that are photographed under different lighting conditions.

Three studies have compared the performance on the Benton Faces test of individuals with WS against that of individuals with DS matched for CA and FSIQ (Bellugi et al., 1992; Rossen et al., 1996; Wang et al., 1995). In each case the WS groups outperformed their controls. However, as already noted, this discrepancy may be primarily due to reduced performance in the DS group, rather than to particularly unusual performance among individuals with WS. If this is the case then this alone does not necessarily indicate preserved face recognition in WS. Nevertheless, in these studies the levels of performance shown by individuals with WS fell within the normal range. Similar results are reported by Bellugi et al. (1988), who found average performance levels across the whole test of 74%, 87% and 80% correct for each of their three individuals with WS. A score of below 70% is considered to indicate defective processing on the Benton Faces; thus these individuals appear to be performing at a normal level. However, it is important to note that the Benton Faces test is designed primarily for use with individuals with neuropsychological deficits. The narrow range of normal performance on the task between 70% and 100% suggests that it is relatively easy for the general population. This raises the possibility that ceiling effects might be present in the standardisation data.

Karmiloff-Smith (1997), assessed 10 individuals with WS with a mean CA of 22;8 years, a mean verbal MA of 10;5 years and a mean performance MA of 6;8 years. In contrast to the studies reviewed thus far, these individuals were matched individually by CA to a control group of 10 TD individuals. Seven members of the WS group scored within the normal range on the Benton Test, two were borderline normal and one individual showed impairment. All of the control group scored within the normal range. However, as detailed scores are not given, there remains the possibility that ceiling effects are present in this control group.

An interview following the task revealed that individuals with WS recognised the faces by specific features such as a cheek bone, or the shape of the nostrils, while the control group talked about the face as a whole. Karmiloff-Smith therefore suggests that the apparently normal levels of performance of the WS participants may be reached by a different means to that of TD controls (see also Wang et al., 1995). Karmiloff-Smith (1997) assessed the same participants in a second face recognition experiment which employed a computerised discrimination task taken from Campbell, Bruce, Import, and Wright (1995). This task requires the individual to discriminate between different faces that vary by a number of different facial elements. These are: facial speech (lip reading), emotional expression, eye gaze direction and the identity of the faces (all features visible, or hairline or eyes masked; Djabri, 1995). Stimuli were presented fullface, or oriented to the right or to the left; some of the faces were very similar to each other while others were very dissimilar. Individuals with WS and controls performed at seemingly comparable levels (over 90% for the TD controls, and over 80% for the WS participants) with the exception of conditions where 'configural processing' was necessary (similar identities, sideways facial orientation, and the masked features conditions). In these conditions, the performance level of the WS group dropped to chance, whilst the control group continued performing at a high level. Karmiloff-Smith attributes this contrast between groups to the type of processing used, suggesting that for individuals with WS to exhibit their characteristically strong face processing skills they need to be able to use facial features rather than processing the global configuration as in typical development. However, Karmiloff-Smith concedes that this is a preliminary study, hence no statistical analysis is provided. Also, given that the interpretations made are based on data where controls are performing at or near ceiling one cannot be entirely certain that typically developing individuals would not find certain conditions particularly difficult if the task was made somewhat harder.

Deruelle, Mancini, Livet, Casse-Perrot and Schonen (1999) employed a similar face matching task to test 12 individuals with WS of mean CA: 11.9 years and mean MA: 5.9 years, matched to two TD control groups, one by CA and one by MA. The WS group performed at the same level as MA controls (accuracy: WS=87.7%, MA=89.3%) and significantly below the CA controls (95.2% accurate) when matching faces by gender, age, gaze direction, emotional expression, and identity across different angles, but at the same level as the CA controls in the lip reading condition. The authors argue that this reflects that matching by lip reading requires featural processing, in contrast to configural processing in the other conditions. However, as above (Karmiloff-Smith, 1997), the results could reflect the fact that ceiling effects are masking a pattern of results in the CA controls which would otherwise be similar to that of the WS group.

Tests of Object Recognition

The Canonical-noncanonical Views Test (Carey & Diamond, 1990) measures the recognition of familiar objects. The ability to name objects from noncanonical or atypical views (such as a teapot viewed from above) is determined from an individual's performance on two subtests, each containing 25 pictures. The first subtest shows objects from noncanonical views. These same 25 objects are then shown from canonical views (e.g., a teapot viewed from the side) to ensure that the individual is familiar with them. A noncanonical views score is calculated as the number of items that were correctly named on the noncanonical subtest as a percentage of those items correctly named on the canonical subtest.

Wang et al. (1995) used this test to compare the performance of their groups of individuals with WS and DS. Similar numbers of canonical views were recognised in both groups (WS: 23.3, DS: 22.7 out of 25), however, the groups differed significantly in their ability to recognise noncanonical views, with the WS group obtaining higher percentage scores (mean: 75.9%) than the DS group (mean: 66.4%). The authors view this as a 'relative strength' in the WS profile, which raises the interesting question of whether the studies reviewed above that purport to show normal abilities in face recognition do so because of a specific or more general strength in recognition memory. However, again the difference between the performance of these groups could equally reflect particularly poor abilities in the DS group.

Hoffman and Landau (2000) report the results of a similar canonical views task. In their study, the performance of 12 children with WS of mean age 11;1 years (range: 7;5 to 15;3) was compared to that of 12 adult controls and 12 TD children who were matched for MA by both verbal and non-verbal ability level on the KBIT and had a mean age of 5;10 years (range: 4;1-7;1). Participants were presented with a computerised task with 4 conditions (canonical view, clear image; canonical view, blurred image; noncanonical view, clear image; and noncanonical view, blurred image) and were required to name 80 objects, 20 from each condition, taken randomly from a pool of 320 images. Results demonstrated that adults performed better than the two groups of children, and that the WS children performed at the same level as the mental aged match controls. The authors suggest that this indicates that "object recognition may be selectively spared."(Hoffman & Landau, 2000). However, the WS group were matched to the controls by both verbal and non-verbal mental age, hence the similarities in the groups' performance indicates that object recognition in WS is at a similar level to their other non-verbal abilities, rather than a spared area of ability.

Tests of Orientation Coding

Atkinson et al., (1997) employed two ‘post-box’ tasks to assess the abilities of 11 children with WS (from a group of 15 WS participants of mean age: 9.7 years, range: 4 to14 years) and a group of 20 TD controls (a subset from a group of 30 TD controls of mean age: 8.1 years ranging from 4 to 20 years). The tasks were taken from the neuropsychological literature and are designed to assess the functioning of the two visual systems, the ventral and dorsal visual streams, responsible for perception and action respectively (Milner & Goodale, 1995). In both tasks, a cylindrical drum is presented. A slot at the front of the drum can be orientated at 0, 45, 90, or 135 degrees. The matching task tests ventral stream functioning: a card is held by a rotatable wooden hand and the individual is asked to rotate the hand so that the card is in the correct position ready to be posted. In the posting task, a test which taps the functions of the dorsal stream, the individual is asked to post the card into the slot. Results indicated that in the matching task the performance of 6 of the individuals with WS was similar to the TD controls, whilst the remaining WS children showed modest deficits. Performance on the posting task was somewhat weaker: 2 individuals with WS displayed errors of a similar magnitude to TD older children and adults, 4 showed errors similar to TD 4 year old controls, whilst 5 made errors that were larger than the control group. The results of this study suggest that individuals with WS are able to match orientations, but experience difficulty if an additional motor action is required.

Orientation matching has also been assessed in WS using the Benton Lines Orientation test (Benton et al., 1978; see Bellugi et al., 1988; Rossen et al., 1996; Wang et al., 1995). In this task the participant is presented with a display of 11 lines oriented 18 degrees apart, and is asked to decide which of these lines matches the orientation of two target lines. Two of Bellugi et al.'s (1988) three children with WS scored at or below 35% correct, while the third child failed the pretest (which requires passing 2 out of 5 practice trials). Wang et al. (1995) report that only 2 of their 10 individuals with WS passed this pre-test. Similarly, Rossen et al. (1996) found that the majority of their 6 participants with WS could not pass the pre-test. These obvious floor effects contrast to the results of Atkinson et al. (1997) above, and tell us only that individuals with WS perform predominantly at a level below that of a ‘severely deficient’ adult, the lowest classification of the test.

Stiers, Willekens, Borghgraef, Fryns, & Vandenbussche (2000) designed a line orientation task, the Pre-school Judgement of Line Orientation task (PJLO), which was set at a more appropriate level for individuals with WS. The task was similar to the Benton lines task, i.e. the individual had to match target lines to a number of choice alternatives. In this task, however, in blocks 1 to 3, one rather than two target lines were presented. Additionally, the number of choice alternatives varied from 2 alternatives in block 1, to 4 in block 2, and 11 alternatives in block 3. Block 4 used items from the original Benton lines task described above, with 2 target lines and 11 response choices. 20 individuals with WS of ages ranging from 5 to 25 years (no mean given) performed at a level, which was slightly below their verbal ability, and at the same level as their non-verbal ability as measured by the WPPSI-R. This highlights the importance of employing tasks which measure the correct range of ability, and appears to suggest, in contrast to the evidence from the standard Benton lines task, and in-line with results of Atkinson et al.'s study, that individuals with WS are able to encode differences in line orientations.

Summary

The visuo-spatial skills of individuals with WS appear to vary considerably between tests. Face recognition is typically seen as a major strength in WS, although the actual evidence for this claim is undermined by the presence of clear ceiling effects among controls or in normative comparison data. Although visuo-spatial abilities are generally impaired in WS, relatively higher levels of performance are seen on some tests, notably the segmented version of the Block Design task, visual search tasks and the Canonical-noncanonical views test. This appears to suggest that individuals with WS are able to process visual information at a global level. Relatively weaker performance is seen on the standard Block Design task, in tests of drawing and in completing drawing versions of the Delis Hierarchical processing task. These difficulties have previously been explained in terms of a local processing bias in WS. This raises a clear contrast; how can individuals with WS show little impairment on such tasks as visual search tasks which require global organisation, given the suggestion (e.g., Bellugi et al., 1988) that they have a preference for processing information at a local level?

The following section attempts to provide an explanation for this apparent contradiction, as well as for some of the differences seen in performance levels across visuo-spatial tasks.

Theoretical implications

Having reviewed the available information regarding visuo-spatial functioning in WS and considered methodological issues in the process, we are now in the position to propose explanations of the results obtained. We have drawn together the whole body of research in the visuo-spatial domain in an effort to make conclusions as to what factors might be constraining performance in WS. The uneven profile of performance of individuals with WS on visuo-spatial tasks may be related to the processing preferences of individuals with WS, such as local or global processing, but additionally it may reflect the specific demands of each task, such as perceptual and constructional requirements.

At present, and as noted above, there is considerable discussion as to whether individuals with WS rely on local methods of processing on visuo-spatial tasks, or whether they use a predominantly global method of processing as in typical development (e.g., Bellugi et al., 1988, Karmiloff-Smith, et al., 1997, Mervis, 1999, Stevens, 1997,Wang et al., 1995, Farran et al., submitted). Bellugi and colleagues argue that individuals with WS show a local processing precedence. Mervis and colleagues propose that they do not have a local bias, but predominantly process information at a global level as in typical development. Pani and colleagues suggest that individuals with WS experience problems in switching from one hierarchical level of processing to another.

Our proposal takes a novel angle by relating the level of performance in WS to the perceptual and constructional demands of the task. We suggest that individuals with WS can perceive information at both global and local levels, as in typical development (as shown by Mervis et al., 1999 and Pani et al., 1999), but that they struggle to use this information to complete visuo-spatial construction at a global level. This constructional process encompasses the abilities needed to perform an overt motor action, the abilities required to perform the internal manipulations of spatial representations necessary for successful motor planning, and the ability to maintain the correct spatial relationships between the parts of an image when reproducing it. Thus, in reference to local and global levels of processing, we suggest that individuals with WS will only show evidence of a local processing bias in tasks with a constructional component. In addition they will not show any particular evidence of a local bias on tasks that are largely perceptual. This account has the potential to reconcile the apparently contradictory evidence that exists in this area.

As already discussed, the poor performance of individuals with WS on the Block Design task has been seen as evidence for a local processing bias (e.g., Bellugi et al., 1988). However, evidence from autism and other areas (e.g., Shah & Frith, 1993) shows that a local processing bias is actually advantageous on this test. A possible solution to this apparent contradiction is provided by Kohs (1923), who designed the Block Design task. Kohs suggests that the task requires “first the breaking up of each design presented into logical units, and second a reasoned manipulation of blocks to reconstruct the original design from separate parts.” Notice that there are two processes involved here. The first – the breaking up of the design – is a perceptual component, and the second – the reconstruction of the design – is constructional. In these terms the evidence suggests that the local processing bias in autism is <u>perceptual</u> (Happé, 1996; Jarrold & Russell, 1997; Shah & Frith, 1993), while the local processing bias in WS is <u>constructional</u> (Bellugi et al., 1988).

Other evidence supports the view that perceptual processing in WS is not particularly driven by a local preference. Individuals with WS show a beneficial effect of segmentation on the Block Design task (Mervis et al., 1999), which would be reduced if stimuli were perceived at the local level. They are also predominantly influenced by the global characteristics of stimuli within visual search tasks (Pani et al., 1999). Performance on the Canonical-noncanonical views task appears to represent a peak in the WS non-verbal profile, and recognition of the noncanonical views is thought to reflect the ability to use local features of an object to recognise the whole form (Carey & Diamond, 1990). This task therefore requires both local and global perceptual processing styles. However, the claim that face processing in WS is influenced by featural analysis (e.g., Karmiloff-Smith, 1997) seems to contradict this proposal. There may be two explanations for this. Firstly, the evidence of a local bias in face processing in WS comes from studies which have employed tasks which may be insensitive to differences in performance levels among controls. Secondly, even if it is shown that face processing in WS is ‘particularly local’, this could reflect the fact that face processing in typical development is seen by some as domain specific process, which is separate from other areas of visuo-spatial cognition (Farah, 1996). If so, then face processing tendencies may be independent of any general visuo-spatial processing style. This would accommodate both the claim for a local perceptual bias when processing faces, and a relatively global bias for perceiving other visuo-spatial information.

A local bias in construction in WS is not only seen on block construction tasks, but also in tests of drawing ability (Bellugi et al., 1988; Bertrand & Mervis, 1996, Bertrand et al., 1997; Wang et al., 1995). Wang et al. explicitly note evidence of this local approach in WS, and observe that their participants with WS were only able to concentrate on the parts of an image one at a time, and did not integrate these to form the global whole. Similarly, Bihrle et al. (1989) and Rossen et al. (1996) concluded from performance on the Delis hierarchical processing task, that individuals with WS approached this task at the local level. However, they reached this conclusion by analysing the quality of an individual’s output, i.e., how they had constructed the image, rather than the quality of the input, i.e. how the image had been perceived. Farran et al. (submitted) and Stevens (1997) demonstrated that these two levels are dissociable in WS by examining both perception and construction of hierarchical figures. In contrast to previous drawing studies, Stevens (1997) only found evidence a predominantly local approach in 5 of his 13 individuals with WS. Nevertheless, when asked to describe the same hierarchical letter stimuli, none of his WS participants showed any evidence of a local processing bias. Similarly, Farran et al. (submitted) report significantly better local than global accuracy in the drawing abilities of their group of WS individuals, which contrasts sharply to the group’s perception of these hierarchical figures where no local processing preference was observed.

Atkinson et al. (in press) looked at the occurrence of sensory visual problems, such as strabismus, reduced stereopsis, or visual acuity loss, in WS. They report that their group of individuals with WS showed no reliable correlation between sensory visual problems, and performance on the three visuo-spatial cognition tasks described earlier. This supports our proposal that visuo-spatial problems in WS are not perceptual.

Pani et al. (1999) suggest that individuals with WS can process at both local and global levels but experience difficulty in switching between these levels of organisation. This differs from our suggestion that the local processing preference in WS is expressed in constructional, but not in perceptual tasks, and is difficult to support for two reasons. Firstly, in the divided attention version of the Delis task, which requires the participant to switch between hierarchical levels, individuals with WS performed at the same level as TD controls (Farran et al., submitted). Secondly, in the Block Design task, a switch is required at the perceptual stage from global perception of the image to the local perception of the individual blocks. This task demand can be eliminated by pre-segmenting the image into its constituent parts (e.g., Mervis et al., 1999). The facilitation effect elicited by pre-segmenting the blocks is equal in WS to that seen in typical development (Farran et al., 2001), suggesting that the switch is equally demanding for both groups. We suggest that faced with the requirements of constructional tasks, individuals with WS experience the normal levels of difficulty at perception, but at construction adopt a piecemeal approach resulting in relative success in reduplicating the local elements of an image, in comparison to the global image. This appears to result from a poor comprehension of spatial relations in WS (Farran & Jarrold, submitted a).

The evidence from Atkinson et al. (1997) indicates a specific problem in performing a motor action in WS. The authors suggest from these results that dorsal stream functioning in WS may be impaired. However, they concede that this deficit can not be the only contributing factor to the pattern of visuo-spatial abilities in WS. Hoffman et al. (in press) indicated, from studying eye movements, that the actual process of construction differed in WS from TD controls. Taken together, the findings of Atkinson et al., Farran and Jarrold (submitted a), and Hoffman et al., support our argument that construction is poor in WS i.e. that the internal manipulation of the individual elements (measured by Hoffman et al.), the maintenance of the spatial representation in reproduction (measured by Farran & Jarrold, submitted a) and the overt motor action required to place each element in the correct position (measured by Atkinson et al.) are problematic processes for the WS population to complete.

The poor ability of individuals with WS to complete the global configuration in block construction tasks is similar to that seen in young children (Akshoomoff & Stiles, 1996; Kramer, Kaplan, Share &, Huckeba, 1999). Kramer et al. (1999) report more broken configurations (defined as any occasion in which the child placed a block outside of the square matrix) produced by children aged 6 to 7 years in comparison to older age groups (8 to 9, 10 to 11, 12 to 13 years). Anecdotal reports suggest that there are a large proportion of broken configurations in WS solutions (e.g., Bellugi et al., 1988). This claim was investigated systematically by Mervis et al., who found that in 76% of solutions, the overall shape was reproduced correctly in WS. The authors state that this is equivalent to the proportion of correct global configurations produced by TD children aged 6 to 8 years as reported by Akshoomoff and Stiles (1996). This implies that the presence of broken configurations in WS solutions on this task could indicate that their level of ability is delayed, rather than deviant. This is supported by the evidence that individuals with WS demonstrate a poor level of ability, but a typical pattern of performance in tasks measuring spatial relations (Farran & Jarrold, submitted a), i.e., delayed rather than deviant abilities..

The development of drawing abilities in children progresses through a local processing stage. At four and a half years, typically developing individuals produce unintegrated drawings much like those produced by nine and a half year old individuals with WS (Bertrand & Mervis, 1996). In addition, the developmental pathway followed by individuals with WS in drawing resembles that seen in typical development (Bertrand et al., 1997). By thirteen and a half years, the drawings of Bertrand and Mervis's WS group had become more organised, and were now equivalent to the drawings of a child of 5 and a half years. Block construction ability in WS also improves during childhood, and by adulthood, level of ability in WS reaches the level of a typically developing 6 year old child (Mervis et al., 1999). As above (Mervis et al., 1999), these results suggest that the local processing bias seen in the drawings and constructional abilities of individuals with WS (e.g., Bellugi et al., 1988, Wang et al., 1995) reflect delayed development. If one were to accept that visuo-spatial output by construction and drawing in WS is at the level of a typically developing child of approximately six years, whilst other skills such as aspects of language are closer to an adolescent's level of performance, can we label these abilities as delayed or deviant? Individuals with WS are aware of their errors, yet still are unable to correct them (Hoffman et al., in press). This contrasts to the performance of a young typically developing child who may fail because they are not sophisticated enough to be aware of their errors, thus errors persist until they simultaneously develop the ability to see and to correct their inaccuracies. Given the huge discrepancy between levels of performance in WS, such a large delay in construction and drawing may in fact be deviant.

The examples above indicate that an individual with WS has problems not so much in the perception of an image, but rather in its reconstruction. Stevens argues that this problem occurs in the planning and execution of a motor action, whilst Pani and colleagues suggest that having processed information at one level, individuals with WS then experience difficulty when a change in the spatial organisation of the image is required. We propose that in order to produce an image through drawing or construction, the individual with WS is forced to rely on a piecemeal method of approaching the task, hence the appearance of a local processing bias at the expense of maintaining the spatial organisation and consequently the loss of the global configuration of the image. The resulting level of output in WS develops until it is the level of a six year old child, at which point individuals with WS reach a ceiling in ability and these abilities cease to develop further.

This piecemeal approach appears to result from difficulty in encoding the spatial relations between the individual elements (Farran & Jarrold, submitted a). As described earlier, spatial relations refer to the relative position of one object to another such as 'above', or 'next to' (categorical spatial relations), or to the precise distance from one object to another (co-ordinate spatial relations) (Kosslyn & Koenig, 1992). Success in construction tasks i.e., mentally deconstructing and manipulating the parts of a model image, and physically reconstructing the image to provide a correct solution, is dependent on accurately preserving the spatial relations between the local elements throughout the construction process. Without this skill or with poor levels of ability, individuals may recreate the local details of the image, but do not place these local elements in the correct relationship to one another, thus the global spatial representation may be incorrect. This appears to be what is observed in the solutions of individuals with WS. In contrast to constructional tasks, the spatial relations between elements is always kept constant in perceptual tasks, and so the ability to understand spatial relations is not a crucial factor for successful task completion. This could explain why perceptual but not constructional abilities might be affected in WS by a lack of comprehension of spatial relations, resulting in a local bias on construction.

Conclusions

A general conclusion that emerges from the review of the above studies is that visuo-spatial abilities in WS are collectively poor, but that the relative level of difficulty of tasks is not consistent across the visuo-spatial domain. We suggest that this variability is partly due to the methodological problems associated with studying WS, but that there are reliable underlying differences in the levels of performance of individuals with WS across a variety of visuo-spatial tests. The evidence from methodologically sound studies has lead us to argue that many of the strengths and weaknesses in WS performance are the result of the perceptual or constructional demands associated with a particular task. Other authors have suggested that individuals with WS might adopt a relatively local approach for all visuo-spatial processing (e.g., Bellugi et al., 1988), yet perceptual ability in WS does not support this hypothesis (e.g., Mervis et al., 1999; Pani et al., 1999). We argue that these accounts highlight that the type of processing preference seen depends on the way in which performance is measured. It appears that individuals with WS show an entirely typical global processing style on tests of perception (see Farran et al., submitted; Mervis et al.1999; Pani et al., 1999), but rely on a local processing approach on tests of construction. Given the need for integration of information (through an accurate comprehension of spatial relations) in constructional tasks, this leads to particular difficulties on these tests, and to a loss of global information.

The methodological difficulties inherent in investigating visuo-spatial abilities in WS, emphasise the need for further studies to confirm whether individuals with WS do have particular problems on tasks which require the construction of a global image. This might be done by further investigations which employ tasks where perceptual and constructional components are dissociable, and by assessing the particular strategies used by individuals with WS to complete these tests. Other areas clearly need further investigation. The area of face processing in WS would be better understood if harder tasks were employed to reduce the occurrence of ceiling effects. The effects of different facial manipulations in tasks such as those used by Karmiloff-Smith (1997) and Deruelle et al. (1999) could then be compared more reliably with that of typically developing individuals. This would indicate whether the apparent local processing preference in the WS group differs from control groups, or whether the difference between groups in previous studies was simply the result of ceiling effects in the controls.

Mental imagery is one area which deserves more attention in WS investigation (see Farran et al., 2001; Farran & Jarrold, submitted b). This includes the ability to pan, translate, rotate, and scan mental images (see Kosslyn, 1994). The processes involved in these transformations and their impact on perception and construction might indicate at which point between apparently intact perceptual abilities and impaired constructional skill, individuals with WS begin to experience difficulty. This could range from storing the image, planning the action at a pre-motor stage, making comparisons during construction between the image being constructed and the to-be-constructed image, to the motor act itself. Greater understanding of these processes would be highly beneficial to the theories currently being developed regarding WS processing.

In summary, our understanding of the unusual pattern of strengths and weaknesses within visuo-spatial cognition in Williams syndrome has given rise to a number of interpretations. In order to evaluate these theories, further investigations that employ both methodologically sound techniques in the areas already tapped, and that examine other as yet untouched areas of visuo-spatial ability are essential.

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Table 1

Mean scaled scores and standard deviations on the subtests of the WISC-R [a,] and WAIS-R [b] across different studies

Study	Mean age (years; months)	n	Picture Completion		Picture Arrangement		Block Design		Object Assembly		Coding	
			Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
Udwin et al. (1987)	11;1	44	3.25	2.18	2.84	2.53	2.14	1.76	3.18	1.82	1.55	1.21
Howlin et al. (1998)	26;6	62	3.39	1.19	3.73	1.77	3.10	1.38	3.30	1.70	2.85	1.11

[a] Udwin et al. (1987), [b] Howlin et al. (1998)