Face and place processing in Williams syndrome: evidence for a dorsal-ventral dissociation

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Individuals with Williams syndrome (WMS) show an interesting dissociation of ability within the visuospatial domain, particularly between face perception and other visuospatial tasks. In this population, using tasks matched for stimuli, required response, and difficulty (for controls) is critical when comparing performance across these areas. We compared WMS individuals with a sample of typically developing 8- and 9-year-old children, and with a sample of adults, closer to the WMS participants in chronological age, in order to investigate performance across two precisely matched perceptual tasks, one assessing face processing and the other assessing proficiency in processing stimuli location. The pattern of performance seen in WMS, but not in controls, implicates a specific deficit of dorsal stream functioning in this syndrome. *NeuroReport* 13:1115–1119 © 2002 Lippincott Williams & Wilkins.

Key words: Dorsal and ventral streams; Face processing; Molecular genetics; Neural systems; Spatial processing; Williams syndrome

INTRODUCTION

Research in typically developing children suggests that face processing undergoes a protracted period of development [1]. Interestingly, from quite early on, individuals with Williams syndrome (WMS) seem to perform better at face processing tasks than at other visual cognitive tasks [2]. However, reports of a dissociation between face and other kinds of visuospatial processing have not equated processing demands across tasks. Studies of face processing typically employ perceptual matching paradigms, while the tasks they are often compared with commonly require visuoconstructional skills. The current study used identical perceptual matching tasks for both experimental tasks, one assessing face processing and the other assessing location processing, thus equating demands across task domains and allowing us to directly assess the issue of a dissociation in WMS between these two cognitive skills.

WMS is a rare disorder that occurs in 1/25 000 births and is caused by a hemizygous deletion encompassing the elastin gene at 7q11.23 and < 20 other genes [3,4]. This syndrome is characterized by distinctive dysmorphologic facial features, mild to moderate mental retardation, distinctive personality characteristics and a unique cognitive profile [5]. A major hallmark of WMS is the dissociation between language (a relative strength) and spatial cognition (a severe and specific impairment) [2,6].

One of the most intriguing aspects of the WMS cognitive profile is a dissociation within the visual processing domain, what Bellugi *et al.* have called a chasm within visuospatial cognition. In drawings and block designs made by individuals with WMS, spatial construction is often severely and specifically impaired. However, these same individuals with WMS show remarkable strength in face processing. This disparity in WMS between spatial construction and face processing, consistently found across different ages, paradigms and samples [7–9], is not seen in individuals with Down syndrome (DNS), who are equally impaired in both. In face processing, not only do WMS perform far better than age and IQ-matched DNS, they have been found to be no different from normal age-matched controls on face recognition tasks [5,10].

This distinction between face and space processing within a specific genetically based population is of great interest, since it may map onto different neural systems in the brain. The primate visual system is subdivided into two anatomically and functionally separate systems [11]. Specifically, the ventral stream (occipito-temporal lobes) is principally involved in processing object properties, whereas the dorsal stream (occipito-parietal lobes) is mainly involved in spatial processes such as object localization and movement detection. Many researchers have examined the functional segregation of these two streams with adults [12,13] and to a lesser degree in children [14,15]. The weight of the evidence to date for the distinction in WMS between face processing and location processing has strongly implicated relative sparing in the ventral stream and a clear deficit in the dorsal stream [16–21].

While there may be relative strength in face processing among individuals in this special population, no study to date has directly quantified the above differences. Most studies of face processing with WMS individuals rely on visuoperceptual tasks and pointing responses, while spatial tasks have been mostly visuoconstructional, such as copying from a model, drawing, block design, etc. These latter spatial tasks involve a significant spatial constructional component, whereas the face identification tasks have minimal motor demands. In the current study we directly confront these limitations of previous research. Instead of contrasting levels of performance on very different neuropsychological measures, we have utilized a paradigm in which the two tasks we use are precisely matched for stimuli and procedures. Further, the face and location tasks were specifically designed to yield comparable performance among adult control participants [13]. With this paradigm, we assess and directly compare face processing skills with proficiency in another strictly visuoperceptual area, processing the location of visually presented stimuli.

The current study compares performance accuracy of a group of WMS participants to that of an adult control group and to a group of mental age-matched children (specifically 8- to 9-year-olds), on a face- and a location-matching task. Findings from an earlier study using this testing protocol with typical adults and children aged 6–12 years showed that although overall accuracy improved with age, performance across the two task conditions was comparable at each age tested [22]. These results demonstrate the effectiveness of the basic task design in balancing face and location processing demands. Given the hypersociability of WMS participants and their tendency to orient preferentially toward faces [23], we wanted to rule out the possibility that any deficits that may be seen on the location-matching task in WMS could be attributed to distraction by the face stimuli themselves. Hence, in addition to the face- and location-matching tasks, a third location control task, using scrambled patterns rather than faces, was also included.

MATERIALS AND METHODS

Participants: Data from 33 individuals diagnosed with WMS aged 12–51 years (mean 27.2; 18 females and 15 males), 19 typically developing right-handed 8- and 9-year old children (mean age 9.0 years; nine females and 10 males) and 24 typical right-handed adults (mean age 20.7 years; 12 females and 12 males) are reported here. The WMS group, recruited as part of an ongoing large program project (P01 HD33113 to UB), received a small monetary sum for participating. They were inducted into the project based on both a clinical diagnosis of WMS and a FISH test (fluorescent *in situ* hybridization) for absence of one copy of the gene for elastin on chromosome 7. Adult controls were college students who were given course credit for participating. Child controls were recruited from local schools, and were given a toy for participating.

In the WMS sample, the mean (± s.d.) WAIS-R Full Scale IQ score was (68.4 ± 8.9). The characteristic disparity among WMS between language and visuospatial ability, also evident in this sample, makes the decision of an appropriate control group a challenging one [24]. Mental age for this WMS sample was calculated using the Peabody Picture Vocabulary Test–III (PPVT-III) [25], and scores ranged from 6.02 to 30.7 years (mean 12.04 ± 4.91). In addition, age equivalent scores from the Beery Developmental Test of Visuomotor Integration, 4th edn (VMI) [26] were calculated, and ranged from 4.1 to 8.0 years (mean 5.78 ± 1.10). Based upon these findings, the WMS group in this study was compared to two different samples of controls, a sample close in approximate mental age: typical children with a mean age of 9 years; and a sample close in chronological age: typical adults with a mean age of 20 years.

Experimental design: Each participant in the study performed the same three tasks, specifically, a face-matching task, a location-matching task and a location control task. The basic design of the three tasks was identical. First, two reference stimuli appeared in sequence in different locations on a computer screen. After a short delay (blank screen), a third (probe) stimulus appeared (Fig. 1). The participant was required to decide whether the probe stimulus matched either of the first two reference stimuli. In the face-matching task, matching was based on identity of the stimulus. In the location tasks, matching was based on location of the stimuli on the screen. In the face-matching and location-matching tasks an identical set of face stimuli were used; thus, these two tasks differed only in the decision the participant was asked to make. In the location control task, matching was also based on location, but the stimuli were scrambled faces. Order of administration for the face-matching and location-matching tasks was counterbalanced across subjects. Because the location control task was created specifically for the WMS population for this study, it was given last to all participants.

The difficulty of any face- or location-matching task can be systematically manipulated by varying similarity of the faces or of the on-screen locations. In the current study, it was important to ensure that the face- and location-matching tasks were comparable in difficulty. To achieve this end, in a preliminary study with typical adults, RT and accuracy scores were collected on a large sample of face- and location-matching trials that varied systematically in difficulty. Items included in our face- and location-matching tasks were selected to yield comparable levels of response time and accuracy performance across the two tasks. Findings from the adult controls in the current study replicated those of the preliminary study. Having anchored the comparability of the basic tasks for adults, we could then test for selective differences in response across the two tasks in our children and in the WMS participants.

Procedure: Figure 1 illustrates the basic procedure for the three tasks in this study. Participants were seated 60 cm from the screen of a Macintosh computer running Psyscope version 1.2 [27]. In all three conditions, the reference and probe stimuli subtended 4.76° visual angle in the vertical dimension and 5.06° in the horizontal. Each type of task was associated with a distinct warning cue displayed to identify the subsequent test trials (green smiley face for the face-matching task and red tic-tac-toe grid for both location tasks). One task block consisted of one warning cue followed by six test trials. Each test trial began with a 500 ms fixation, followed by a 1000 ms interstimulus interval (ISI). Next, the two reference stimuli appeared in sequence for 500 ms each, separated by a 250 ms ISI, and then, after a 500 ms delay, the probe stimulus was presented. During presentation of the probe, participants made a button press (yes or no) indicating whether the probe stimulus matched either of the two reference stimuli with respect to the attended property (identity or location). Adult controls were

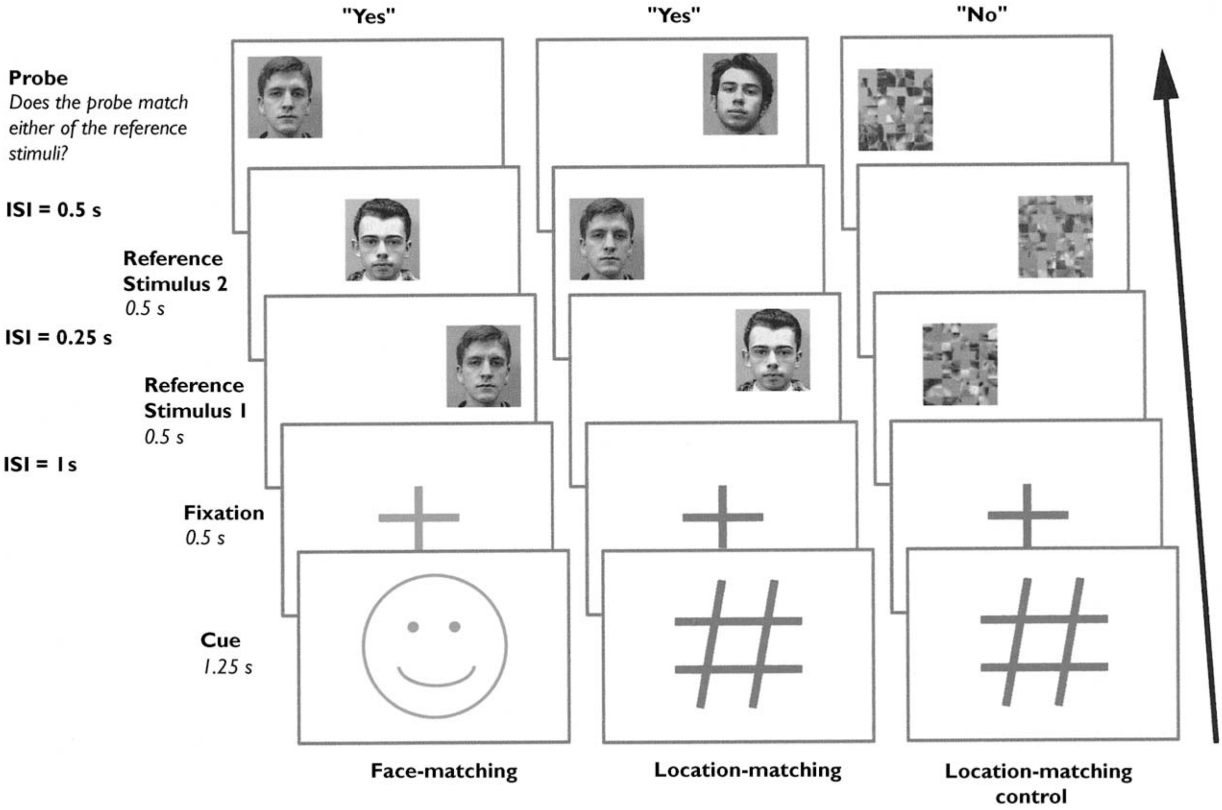

Fig. 1. Diagram of task design and sample stimuli. For all tasks, a warning cue appeared at the beginning of a block of six trials. The green smiley face in the face-matching task cued the participant to attend to the identity of the stimuli. The red grid icon in the location tasks cued the participant to attend to the location of the stimuli. Stimuli could appear in 12 possible locations. An inter-trial interval of 0.5 s separated one trial from the next in all three tasks.

allowed a maximum of 2000 ms to respond, while child controls were allowed 2500 ms and WMS 3500 ms. It is likely that this difference did not truly alter the tasks since both control groups responded well within their allotted times.

Stimuli used in both the face-matching and location-matching tasks were black and white photographs of male faces. For the location control task, scrambled black and white patterns were used. These scrambled stimuli contained the same range of spatial frequencies as the regular face photographs, but were not recognizable as faces. For all three tasks, half the trials contained matching probes and half did not. During the face-matching task, reference and probe stimuli locations never matched; during location tasks, reference and probe stimuli never matched in identity. For the control adults and children, 48 trials per task were given (eight blocks). To maximize performance among the WMS, shortened versions of the three tasks were used. They consisted of 18 trials each, identical to the first 18 trials of each task given to controls. To take into account the difference in task lengths across groups (48 versus 18 trials per task), the analyses reported here were also performed using control data from only the first 18 trials per task; the results did not change.

The WMS and child control groups were given a minimum of eight practice trials per task and the adult group, four practice trials. During the practice and experimental tasks, typical adults and children were provided with feedback (a computer beep) for incorrect answers. WMS participants were given feedback only during their practice trials in order to minimize distractions, since WMS individuals tend to be hypersensitive to sounds [28].

Participants were excluded if their performance was below chance on all three tasks (four out of the original sample of 37 WMS). With 48 trials, chance performance for controls was ~ 0.59 ($p = 0.05$) or below according to the binomial probability distribution (note: given that WMS had fewer trials, in order to maintain a comparable range of scores across the study groups, WMS were retained with performance of 0.67 ($p = 0.07$) or higher. All but two WMS scored well above the 0.05 chance level for 18 trials on at least one task). Mean substitution (of the appropriate participant group mean) was used to deal with missing data points; this was performed on only one cell (one WMS participant on the location control task). Because WMS is such a rare clinical group, that group was not restricted by handedness. Five non-right-handed WMS individuals were included in these analyses. When the analyses were rerun excluding these five subjects, the results did not change.

RESULTS

Accuracy was investigated with a $3 \times 2 \times 2 \times 2$ (group $\times$ order $\times$ gender $\times$ task) mixed design ANOVA. Group (WMS, adult controls, child controls), order (face-matching first, location-matching first) and gender were between subject variables. Task (face-matching, location-matching, location control) was the within subject variable. Preliminary analyses revealed that order and gender were not involved in any of the significant effects, and were dropped from further analyses. In the resulting 3×2 (group $\times$ task) ANOVA, the groups $\times$ task interaction was significant ($F(4,146) = 16.52$, $p < 0.001$). Analysis of simple effects (by group) indicated that performance across the three tasks differed significantly only in the WMS group ($F(2,64) = 31.21$, $p < 0.001$; Fig. 2). Bonferroni's correction was applied to correct for multiple comparisons. A follow-up Tukey HSD test for all pairwise comparisons revealed that WMS performance on face-matching was significantly better than performance on both the location-matching ($p < 0.001$) and the location control tasks ($p < 0.001$). Performance on the two location tasks was not different ($p > 0.6$). This pattern of performance is striking and uniform across subjects; 30 of the 33 WMS participants show the pattern regardless of age and overall ability level. Performance in the adult and child control groups did not differ across tasks.

DISCUSSION

The findings from this study document a striking disparity between face and location processing in a large sample of WMS individuals under experimental conditions in which processing demands were carefully balanced. The findings (Fig. 2) are consistent with results from previous studies on face and location processing in WMS and confirm the spatial deficit even in the absence of a motor component [2]. One possible account of the WMS poor performance on the location-matching task in this study could have been that because the stimuli in the location-matching task were always faces, WMS were unable to repress face processing and were simply off task. By this account, the disparity in performance observed between the face- and location-matching tasks could reflect a strong attraction to face stimuli rather than deficits in location processing. Three findings argue against this account. First, if WMS individuals were in fact engaged in face matching during location-matching trials, their accuracy scores should have been substantially lower. Recall that during location-matching, the reference and probe faces never matched, thus if WMS individuals attempted to match faces during these trials, all of their responses should have been "no" and their performance should have been at 50%. In fact, their performance during location matching was substantially higher than 50%. Further, examination of individual patterns of responses showed a comparatively even distribution of yes and no responses for WMS participants during location matching, suggesting that they were not simply defaulting to a face matching strategy. Second, the performance of the WMS group on the location control task was virtually identical to that on the location-matching task. Thus, even when no distracting face stimuli were present, location processing performance was still impaired. Lastly, the selective impairment of location processing cannot be characterized as an immature processing strategy. In a study of typically developing 6–12 year olds using tasks similar to those reported here, but with increased processing demands, we demonstrated that the increase in difficulty led to a selective decline in face, but not location processing [22]. Thus, the performance profile for the WMS represents a pattern of selective deficit rather than developmental delay.

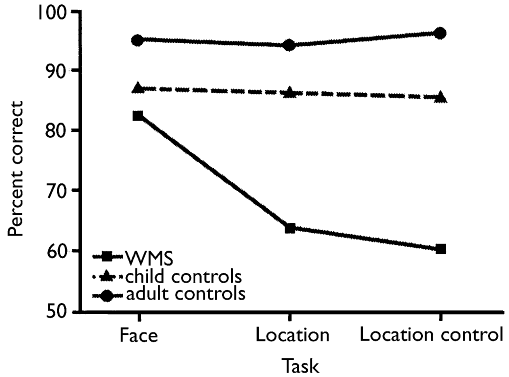

Fig. 2. Task performance across groups. It appears that WMS participants and controls close in mental age performed at similar levels of accuracy on the face-matching task (82.2% for WMS *vs* 86% for child controls), while their performance levels on the location-matching task were markedly different (62.7% for WMS *vs* 85.6% for child controls).

CONCLUSIONS

The major finding of this study confirms previous reports of strong face processing in the WMS population, as well as reports of a spatial deficit. However, it extends the documentation of poor performance in the spatial domain to include a perceptually based location-processing task matched precisely in stimulus and response demands to the face-processing task. The findings from this study thus highlight in a new and more compelling way the contrast between face processing and spatial processing abilities in the WMS group. Our findings serve to confirm the existence of a chasm within visuospatial cognition, as well as to characterize it in greater depth. The current findings add further support to claims of a functional dissociation in WMS which is reflected in contrasting patterns of strength and deficit in processes mediated by different visual processing streams. Specifically, WMS appear to manifest deficits in processes associated with the dorsal stream, while exhibiting relative strengths in processes, such as face processing, that are mediated by the ventral stream. The current findings are consistent with the hypothesis of a dissociation in dorsal versus ventral stream processing that is beginning to gain support from neurophysiological, neuromorphological and cellular studies of WMS underlying brain systems.

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