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People with Williams syndrome process faces holistically

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Abstract

This study compared the performance of 47 adolescents and adults with Williams syndrome to 39 age-matched controls on a face recognition task. Using the whole–part paradigm developed by Tanaka and his colleagues, we found that although performance overall was lower in the participants with Williams syndrome, both groups showed similar patterns of performance across the different conditions. Both groups performed significantly better in the whole-face than in the isolated-part test condition for upright faces, but not for inverted faces. The whole-face advantage only in the upright condition provides strong evidence that people with Williams syndrome encode and recognize faces holistically in the same way as normal controls, suggesting the use of similar underlying neurocognitive mechanisms. These findings contradict earlier reports in the literature that people with Williams syndrome process faces abnormally.

Keywords: Williams syndrome; Face recognition; Holistically

1. Introduction

Williams syndrome (WMS) is a genetically based neurodevelopmental disorder that has captured the interest of cognitive neuroscientists because of the striking and unusual pattern of cognitive abilities that is characteristic of individuals with this syndrome (e.g. Bellugi, Marks, Bihrle, & Sabo, 1988; Karmiloff-Smith et al., 1997; Mervis, Morris,

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Bertrand, & Robinson, 1999). Relative strengths in auditory short-term memory and language coupled with profound deficits in visual-spatial constructive skills define the cognitive phenotype (Bellugi, Wang, & Jernigan, 1994; Mervis et al., 2000; Morris & Mervis, 1999). Although overall cognitive level is in the moderate range of mental retardation, within the visual-spatial domain face recognition skills appear to be relatively preserved in WMS. On standardized tests including the Benton Test of Facial Recognition and the Rivermead Behavioral Memory Test, people with WMS generally perform within the normal range, and better than mental-age-matched controls (Bellugi et al., 1988; Karmiloff-Smith, 1997; Udwin & Yule, 1991). Despite their strengths in recognizing faces, numerous researchers claim that people with WMS do not process faces normally (Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999; Elgar & Campbell, 2001; Gagliardi et al., 2003; Karmiloff-Smith, 1997; Karmiloff-Smith, Scerif, & Thomas, 2002). The goal of this study was to address this apparent paradox regarding face processing skills in WMS: is it possible to be good at recognizing faces using abnormal processing mechanisms?

There is general agreement that our ability to recognize and discriminate faces so easily represents one of the most remarkable human skills and depends on specific cognitive and neural mechanisms (e.g. Farah, Wilson, Drain, & Tanaka, 1998; Haxby, Hoffman, & Gobbini, 2002). To a much greater degree than most other objects, faces are encoded holistically rather than by local features (Bradshaw & Wallace, 1971; Farah, 1996). Specifically, faces are encoded in terms of a template-like representation of the whole rather than simply in terms of their component parts. It has been shown that this mechanism typically breaks down when faces are presented in inverted orientation, as inversion is thought to disrupt the expected configuration of the face (Rhodes, Brake, & Atkinson, 1993; Valentine, 1988). Although children's performance on face processing tasks improves with age, there is evidence using inverted faces and other paradigms that even young children process faces holistically (Baenniger, 1994; Carey & Diamond, 1994; Freire & Lee, 2001).

Tanaka and his colleagues introduced the whole–part method, which was specifically designed to operationalize the distinction between holistic and feature-based face processing (Tanaka & Farah, 1993; Tanaka, Kay, Grinnell, Stansfield, & Szechter, 1998). In the whole–part procedure, a sample upright face stimulus is briefly exposed, followed by two types of recognition tests. In the whole-face test condition, the sample face and a foil face differing from the sample by only one feature (eyes, nose, or mouth) are presented. Alternately, in the isolated-part test condition, one feature from the sample face and the corresponding foil feature are presented. The logic of the whole–part method is that if upright faces are processed holistically, the individual features of a face will be recognized more easily in the context of the whole face in which they were learned than in isolation, but that any holistic processing advantage would not be operative for whole and part test stimuli presented in inverted orientation. Using this method, Tanaka and his colleagues have found that adults and children as young as 6 years old exhibit a whole-face test advantage for upright but not inverted faces. Joseph and Tanaka 2003 successfully adapted the whole–part method to investigate face recognition in children with autism. They found that children with autism showed a holistic processing advantage when face recognition depended on mouths, but were significantly impaired, compared to matched controls, in recognizing the eye region of the face. Their study demonstrated that the whole–part method is especially suited to identifying normal and aberrant face processing mechanisms.

As noted above, people with WMS perform within the normal range of performance for age on standardized tests of face recognition, classification, and memory (Bellugi, Bihrle, Neville, Jernigan, & Doherty, 1992; Bellugi et al., 1994; Karmiloff-Smith, 1997; Udwin & Yule, 1991). However, it has been suggested by a number of researchers that they do not process faces holistically. For example, in their recent review of the literature on face recognition in developmental disorders, Elgar and Campbell argue, "relatively good performance on face discrimination tasks in Williams syndrome is achieved via a piecemeal route, as in individuals with autism" (Elgar & Campbell, 2001, p. 709). And Karmiloff-Smith concludes that "Williams syndrome face processing is not intact and it develops differently" (Karmiloff-Smith, 1997, p. 522).

These claims of aberrant face processing in WMS are based on two studies. Karmiloff-Smith (1997) gave a group of ten adolescents and adults with WMS and ten age-matched controls the Benton test. After the test was administered in the standard way, she then presented some of the test faces in inverted orientation, and questioned the participants about how they remembered the faces. In a follow-up experiment with the same participants, Karmiloff-Smith administered a computerized face processing test battery developed by Campbell and her colleagues for children aged 4–10 years. Although no data or statistics were presented, Karmiloff-Smith reported that the participants with WMS were less disturbed than controls when faces were presented in inverted orientation and that they pointed to features when asked how they remembered a face. Karmiloff-Smith also reported that item analyses for the second experiment suggested that the participants with WMS were at chance on test items that required configural processing. It is difficult to evaluate this study since few methodological details were presented, and the informal nature of the procedures used and analyses conducted do not allow one to objectively evaluate the conclusions. Karmiloff-Smith (1997) acknowledged the preliminary nature of her study, which included a very small number of participants with WMS who varied widely in both age and IQ.

A second experimental study on face recognition in WMS was conducted by Deruelle et al. (1999; Experiment 2). They compared 12 participants with WMS between the ages of 7 and 23 to two control groups, one matched on age and the other matched on mental age, on a same–different discrimination task in which faces and houses were presented in upright or inverted orientation. All the groups made relatively more errors on the inverted faces than on the houses, and for the control groups this pattern was confirmed by a significant interaction between stimulus type and orientation. This interaction did not reach statistical significance for the WMS participants. However, the data for the WMS group (Deruelle et al., 1999, Table 2, p. 287) show that the mean difference in errors on the inverted faces compared to upright faces (1.23) was higher than on inverted houses (0.43), and this difference on faces was actually higher for the WMS group than the age-matched controls (0.83). Thus, the data obtained from this study were in the direction predicted if the participants were using a holistic processing mechanism. The lack of a statistically significant effect is not surprising given the small number of participants, who varied widely in age and ability level. It is doubtful that there was sufficient statistical power to detect a significant effect from this group of participants. Nevertheless, the authors conclude from this experiment that their participants with WMS "are incapable of encoding faces in terms of configurational information and encode both upright and inverted faces through local characteristics" (Deruelle et al., 1999, p. 288). Taken together, neither of these studies on face recognition in WMS provide compelling support for the view that people with WMS process faces in an aberrant or atypical way.

In light of the evidence from standardized data, which suggests that face processing is a relative strength in WMS, we were surprised by the claims that in people with WMS face recognition depends on abnormal mechanisms that process featural information (Elgar & Campbell, 2001; Gagliardi et al., 2003; Karmiloff-Smith et al., 2002). Our study was designed to explore the question of how people with WMS recognize faces using a more rigorous and sensitive methodology. We address the methodological weaknesses of earlier work by directly testing whether people with WMS process faces relying primarily on holistic or featural mechanisms. We included a relatively large group of adolescents and adults with WMS, and used the whole–part method developed by Tanaka and his colleagues (Tanaka & Farah, 1993). In this paradigm, accuracy differences between the whole- and part-face conditions can be unambiguously interpreted as evidence for the use of holistic or featural processing mechanisms in face recognition. We used the same stimuli and similar procedures as those developed by Joseph and Tanaka 2003 because they had been used successfully to investigate face processing in individuals with autism, a different developmental disorder with which WMS has been compared (cf. Elgar & Campbell, 2001; Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995).

2. Methods

2.1. Participants

Participants in this study included 47 adolescents and adults with WMS (28 females and 19 males) ranging in age from 12 to 36 years old, who were recruited through the Williams Syndrome Association. The diagnosis of WMS was confirmed for all participants by a geneticist and the FISH test. Four additional adolescents with WMS were excluded from the analyses due to their inability to focus and complete the task, or due to a clear bias demonstrated by always pushing the same right-side button throughout the experiment in making their decision. A total of 81 normal controls between the ages of 12 and 37 were tested, however 42 were excluded from the final sample because their IQ scores were more than one standard deviation above or below the mean. The remaining 39 controls (25 females and 14 males) were well-matched on age to the participants with WMS ($t(84) = 0.89$, NS).

All participants received the Kaufman Brief Intelligence Test (KBIT; Kaufman & Kaufman, 1990) and the Peabody Picture Vocabulary Test-III (PPVT-III; Dunn & Dunn, 1997) to assess IQ and verbal knowledge. Basic visual-spatial abilities were measured using the Block Design subtest from the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999). Participants were also administered the short form of the Benton Test of Facial Recognition (Benton, Hamsher, Varney, & Spreen, 1983). Table 1 presents

Table 1
Participant characteristics

	Williams syndrome ($n = 47$)			Normal controls ($n = 39$)		
	M	(SD)	Range	M	(SD)	Range
Chronological age	20;10	(7;3)	12;1–36;2	19;5	(6;8)	12;1–34;5
Full Scale IQ (KBIT)*	68.91	(12.2)	51–100	106.15	(6.6)	91–115
Verbal IQ (KBIT)*	75.55	(11.3)	41–102	106.10	(7.2)	87–119
Nonverbal IQ (KBIT)*	67.98	(14.1)	40–98	105.21	(7.3)	87–119
Vocabulary (PPVT-III)*	79.78	(9.9)	51–103	110.25	(12.0)	81–136
Benton facial recognition**	21.6	(2.6)	16–27	22.9	(1.9)	18–27
Block design (T score; $M = 50$)*	26.78	(3.8)	21–35	55.20	(10.5)	33–70

Groups significantly different at $*P < 0.001$, $**P < 0.02$.

the characteristics of the participants on these standardized measures. There was a significant difference between the groups on the Benton test ($t(84) = 2.51$, $P < 0.02$, $\eta^2 = 0.07$). In the WMS group eight participants (17.1%) had Benton scores in the borderline and below average range, while only two (5.2%) of the controls obtained below average Benton scores. As expected, compared to the controls, the participants with WMS had significantly lower IQ scores ($t(84) = 17.03$, $P < 0.0001$), PPVT-III standard scores ($t(84) = 12.92$, $P < 0.0001$), and Block Design scores ($t(78) = 16.24$, $P < 0.0001$).

2.2. Materials

Face stimuli were the same as those used by Joseph and Tanaka 2003, which were constructed from grayscale, digital face photos of school-age children posing with a neutral expression. Adobe Photoshop 5.0 software was used to make 12 target faces (six boys and six girls) from the face outline of one child, the eyes of a second child, the nose of a third child, and the mouth of a fourth child. Three foil faces were made for each of the 12 target faces by replacing either the eyes, nose, or mouth of the target face with the corresponding feature from an unused photo. Composite whole faces were used to ensure that target and foil faces were equated for naturalness. The whole-face stimuli were cropped to 3 inches in width and approximately 4 inches in length. Part-face stimuli were made by cropping each feature (eyes, nose, mouth) in Photoshop so as to maintain the original position of each feature on the canvas. Fig. 1 shows sample whole- and part-face stimuli. In addition to the face stimuli, a set of whole–part training stimuli was made from grayscale graphic images of common objects.

2.3. Procedure

Participants were tested individually. Stimuli were presented on a 15 inch Dell Inspiron laptop computer using SuperLab Pro 2.0 software. Responses were recorded with two buttons from a 4-button Cedrus 410 button box. All participants underwent a training procedure with non-face whole–part stimuli to ensure that they understood the test format and appropriate use of the response buttons. Training included a series of trials in which a

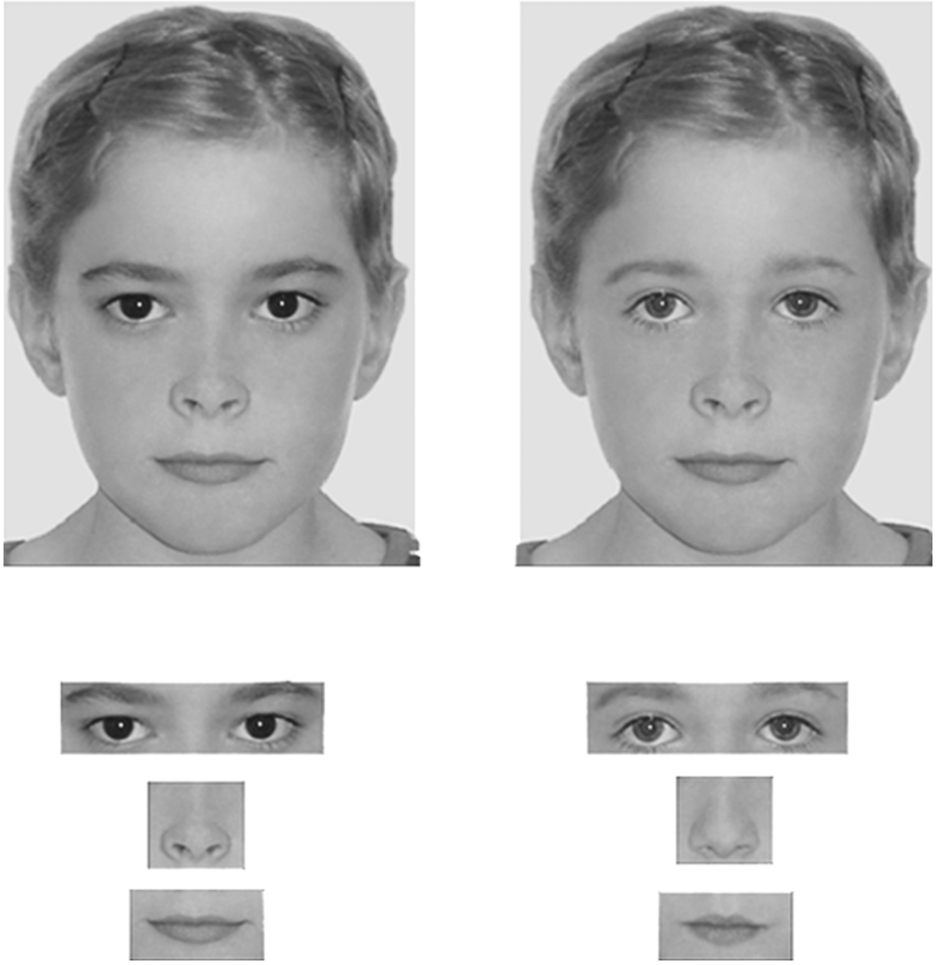

Fig. 1. Sample whole and part test stimuli.

line drawing of an object appeared for inspection (e.g. a flag or a boat), followed by the presentation of a target–foil pair consisting of the original drawing and a similar but not identical line drawing from the same object category (e.g. two flags). The training stimuli were manipulated as described below for the test stimuli (i.e. whole/part and upright/inverted presentation). Feedback was given during training to ensure that the participants understood the match-to-sample procedure.

Following the training procedure, participants were asked to look carefully at each face that would be presented. For each trial, a whole-face stimulus was presented for 3.5 seconds, followed immediately by a two-choice recognition test. In the whole-face test condition, participants saw the original target face alongside a foil face that differed by one feature. In the isolated-part test condition, only the feature on which the target and foil faces differed was presented. Participants were instructed to select the face or face part that best matched the face they had just seen by pressing the button (left or right) corresponding to the location of their choice on the screen. Target and foil face stimuli remained on the screen for up to 8 seconds and disappeared as soon as a button-box response was made, followed by 1 second of a blank screen before the presentation of the next target face stimulus.

Six upright and six inverted faces were presented in the whole-face and isolated-part test conditions for each of the three features, resulting in a total of 72 trials. In the inverted condition, whole target faces were presented in inverted orientation for a 3.5 second exposure period, and stimuli in the subsequent forced-choice test phase were also presented in inverted orientation. The set of six faces (three boys and three girls) that were presented in the upright condition for half the participants were presented in the inverted condition for the other half, and vice versa. The order of stimulus presentation was randomized for *test type* (whole, part), *orientation* (upright, inverted), and *feature* (eyes, nose, mouth), with the constraints that the same target face was never tested over consecutive trials and the same feature was never tested over more than two consecutive trials. The position of correct response was counterbalanced. Testing was administered in three blocks of 24 trials. Participants were given the opportunity to rest between blocks.

3. Results

Table 2 presents the mean number and percentages of correct responses for each group in each condition. Preliminary analyses of the data revealed no significant effects of sex or age of participants and were therefore not included in later analyses. A mixed model ANOVA with group as the between subjects factor and orientation (upright vs. inverted), test type (whole vs. part), and feature (eyes vs. nose vs. mouth) as repeated measures factors was conducted on the number of correct responses. Although overall accuracy was

Table 2
Correct responses for each feature in each condition

	Williams syndrome ($n = 47$)			Normal controls ($n = 39$)		
	Eyes	Mouth	Nose	Eyes	Mouth	Nose
Upright whole						
M (SD)[a]	4.6 (1.4)	4.5 (1.3)	3.8 (1.2)	5.3 (0.93)	5.2 (0.98)	4.4 (1.2)
Percentage	76	75	63	89	87	73
Upright part						
M (SD)	4.0 (1.2)	3.6 (1.4)	3.4 (1.3)	5.3 (0.91)	4.3 (1.4)	3.8 (1.3)
Percentage	67	61	57	88	72	63
Inverted whole						
M (SD)	3.0 (1.3)	3.4 (1.4)	3.2 (1.3)	3.9 (1.2)	3.7 (1.2)	3.7 (1.3)
Percentage	50	57	55	66	62	62
Inverted part						
M (SD)	3.2 (1.5)	3.4 (1.3)	2.9 (1.3)	3.9 (1)	4.1 (1.2)	3.8 (1.3)
Percentage	52	57	49	65	68	63

[a] Means and percentages are for six trials.

better in the control group than the WMS group ($F(1,84) = 38.45$, $P < 0.001$), the two groups showed a similar pattern of performance, as indicated by the lack of significant interactions between group and any of the repeated measures.

To further examine the specific patterns on performance within each group, repeated measures ANOVAs with orientation (upright vs. inverted), test type (whole vs. part), and feature (eyes vs. nose vs. mouth) as within subjects factors were conducted for each group separately. Of main interest was the presence of a significant interaction between orientation and test type for each group (WMS group, $F(1,46) = 6.67$, $P < 0.02$; control group, $F(1,38) = 8.12$, $P < 0.01$). Analysis of this interaction indicated that both groups were more accurate in the whole-face than in the isolated-part test condition for upright faces, but not for inverted faces. A whole-face advantage in the upright but not the inverted condition is expected to result when faces are processed holistically. Thus, when faces were presented in the upright orientation, the participants with WMS performed significantly better in the whole-face than in the isolated-part condition (71.3% vs. 61.5% correct, respectively) ($t(46) = 3.89$, $P < 0.001$), as did the normal controls (82.9% vs. 74.1% correct, respectively) ($t(38) = 3.22$, $P < 0.01$). There was no significant difference in accuracy between the whole-face and isolated-part condition for either group when stimuli were inverted.

As shown in Fig. 2, a strong 'inversion effect' was also observed in the performance of both groups in the whole-face condition, as indicated by a significant main effect of orientation ($F(1,46) = 72.32$, $P < 0.0001$ for the WMS group; $F(1,38) = 79.09$, $P < 0.0001$ for the control group). Pairwise comparisons showed that performance was significantly impaired when whole-face stimuli were presented upside-down, for both groups (71.3% correct in upright vs. 54% in inverted presentation for the WMS group, $t(46) = 8.5$, $P < 0.001$; 82.8% correct in upright vs. 62.9% correct in inverted

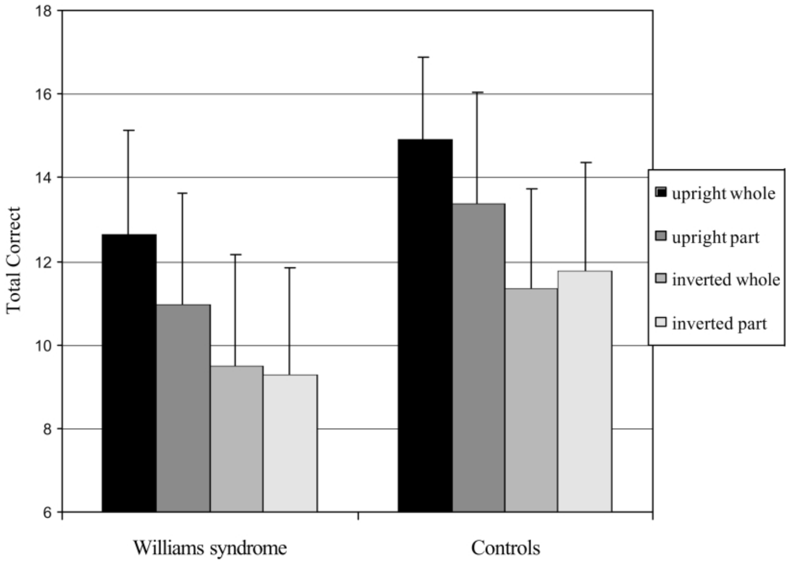

Fig. 2. Number of correct responses as a function of orientation and test type.

presentation for the controls, $t(38) = 8.89$, $P < 0.0001$). The inversion effect is considered to be an indirect measure of holistic processing, in addition to the more direct whole-face advantage results found for both groups when presented with upright stimuli.

Differences in accuracy were found for both groups as a function of the face feature that recognition depended on, specifically for faces presented in upright orientation (see Table 2). This was confirmed by analyzing the significant interaction between orientation and feature that was obtained in the ANOVAs conducted separately for each group, described above. For the controls, in the upright condition, the orientation by feature interaction was highly significant ($F(2,37) = 9.37$, $P < 0.001$), while for the WMS group the interaction approached significance ($F(2,45) = 4.051$, $P < 0.06$). The participants with WMS performed significantly better when recognition depended on the eyes than when it depended on the nose ($t(46) = 2.9$, $P < 0.001$), and when recognition depended on the mouth compared to the nose ($t(46) = 2.96$, $P < 0.04$). For controls, accuracy was consistently better when recognition depended on the eyes than when it depended on the other two features tested (the nose, $t(38) = 7.38$, $P < 0.001$; the mouth, $t(38) = 3.12$, $P < 0.001$), as well as when recognition depended on the mouth compared to the nose ($t(38) = 3.9$, $P < 0.001$) for stimuli presented in upright orientation (see Table 2). As expected, there were no significant differences in accuracy between features when stimuli were presented in inverted orientation for either group.

In order to investigate possible relationships between face processing accuracy and other cognitive abilities or age we computed correlations between standardized and experimental measures. These analyses revealed no significant correlations for the WMS group between scores on the experimental measures of face processing and age, IQ, or performance on the block design test. However, scores on the Benton test of facial recognition were positively correlated with overall accuracy ($r(45) = 0.44$, $P < 0.01$), and with performance in the whole-face condition for upright faces ($r(45) = 0.43$, $P < 0.01$). A similar pattern of correlations was found for the control group. Thus, the standardized measures of cognitive functioning or age were not correlated significantly with the experimental face processing measures. However, moderate positive correlations were found for the Benton test score and face processing performance in the upright condition ($r(37) = 0.36$, $P < 0.05$), especially when recognition depended on the eyes ($r(37) = 0.38$, $P < 0.02$).

4. Discussion

This study was designed to test whether people with WMS process faces holistically, using the same mechanisms that have been identified for children and adults without disorders (Farah et al., 1998; Haxby et al., 2002). Using the part–whole paradigm developed by Tanaka and colleagues (e.g. Tanaka & Farah, 1993), we found that adolescents and adults with WMS showed the whole-face advantage for upright faces but not inverted faces, a pattern that held for all the face features that were tested. This pattern of findings provides strong evidence in support of the view that people with WMS process faces normally. Furthermore, the significant correlations obtained between performance on the experimental task and the Benton standardized test of face recognition suggest that people with WMS are skilled in recognizing faces because they do so using specialized mechanisms that depend on holistic processing of faces.

These findings contradict earlier reports in the literature that people with WMS process faces abnormally, relying on local features rather than via holistic representations (Deruelle et al., 1999; Karmiloff-Smith, 1997). As noted in Section 1, claims about aberrant face processing in WMS were based on studies that were methodologically weak, and did not depend on paradigms specifically designed to test whether participants process faces holistically. Despite the limitations of these studies, there is a widespread view that face processing is abnormal in WMS, and develops in completely different ways (Elgar & Campbell, 2001; Gagliardi et al., 2003; Karmiloff-Smith et al., 2002).

Similar claims have been made about some other cognitive systems in WMS, including language, theory of mind, and visual-spatial cognition (e.g. Bellugi et al., 1988; Bihrle, Bellugi, Delis, & Marks, 1989; Karmiloff-Smith et al., 1995; Pinker, 1994). Early reports about the remarkable linguistic abilities of people with WMS have been tempered by more recent studies suggesting that their grammatical skills are commensurate with their cognitive level, not superior (e.g. Mervis et al., 1999). Despite claims to the contrary (Karmiloff-Smith et al., 2002), there is no evidence that children with WMS acquire language any differently than other children, although they may be delayed in the onset of first words and phrases, as would be expected given their mental retardation (Morris & Mervis, 1999). Similarly, in the domain of theory of mind one published report concluded that WMS involves an “islet of relatively preserved ability” (Karmiloff-Smith et al., 1995, p. 202). However, more recent studies, which included appropriately matched control groups, found that children and adolescents with WMS were no different from others with mental retardation on standard tasks tapping false belief and related concepts (Sullivan & Tager-Flusberg, 1999; Tager-Flusberg & Sullivan, 2000).

In contrast to language, theory of mind, and face recognition, children and adults show profound impairments on tasks tapping visual-spatial abilities (Bellugi et al., 1988, 1992, 1994; Mervis et al., 1999). It has been suggested that people with WMS are so impaired on visual-spatial tasks because they process spatial information in an abnormal way. Based on their performance on drawing and other constructive visual-spatial tasks, Bellugi and her colleagues argued that instead of attending to both global and local aspects of a spatial display, people with WMS attend exclusively to the local individual elements, and ignore the overall global arrangement (e.g. Bellugi et al., 1992), echoing the claims made about atypical face processing (e.g. Elgar & Campbell, 2001). However, using a paradigm that is highly sensitive to global processing of spatial arrays, Pani, Mervis, and Robinson (1999) found that adults with WMS show the same pattern of performance and response times as normal controls, a pattern that confirmed their ability to attend to the global characteristics of a stimulus arrangement. More recently, Hoffman, Landau, and Pagani (in press) analyzed how children with WMS solve complex visual-spatial tasks using eye fixation and error patterns. The children with WMS were significantly impaired on the complex tasks; however, this was because they relied on inefficient executive processes and impoverished spatial representations but there was no evidence for abnormal organization of cognitive mechanisms.

Thus, across a range of research studies and cognitive systems there have been numerous contradictory claims made about WMS by researchers from widely different theoretical camps, many of which have not held up after more systematic investigation. Given the strong interest in WMS by the fields of developmental and cognitive psychology, it would seem important to hold researchers investigating cognition in WMS to the same standards as other cognitive scientists. Too often, studies involve very small numbers of participants with WMS who vary widely in age and IQ level, and appropriate comparison groups have not been included. Furthermore, many studies have not employed experimental tasks or paradigms drawn from contemporary cognitive science that are most sensitive to elucidating the mechanisms that underlie performance in a particular cognitive domain, and which are suitable for use with a population that has mild to moderate mental retardation. Research on WMS has much to inform us about how cognitive systems are organized, but can only do so if such research is carried out in a methodologically rigorous and appropriate way.

In this paper, we presented evidence that adolescents and adults with WMS process faces holistically, and not in an aberrant part-based way. Our evidence is based on the *pattern* of performance across different conditions included in our experiment (see Fig. 2). Specifically, participants with WMS and normal controls were better able to recognize upright face features when presented in the whole-face condition compared to when individual features were presented, but this whole-face advantage disappeared when the face stimuli were presented in the inverted condition. Furthermore, both groups performed best on eyes, although this did not reach significance for the group with WMS. This pattern of performance contrasts sharply with the performance of young adolescents with autism who show evidence for holistic face processing only when recognition depends on the mouth, and show aberrant and impaired processing of eyes compared to other face regions (Joseph & Tanaka, 2003). These data, based on the same stimuli and paradigm used in our study, suggest that people with autism process faces quite differently from people with WMS, contradicting the conclusions drawn by Elgar and Campbell (2001).

Although the participants with WMS showed similar patterns of performance across the different conditions in the experiment, their overall level of performance was significantly lower than the age-matched controls. We attribute this to general cognitive impairments that are characteristic of WMS, as most, but not all, of our participants with WMS had IQ scores in the mild to moderate range of retardation. Although the majority of the WMS participants performed within the normal range on the Benton, our experimental task was significantly more challenging (see Fig. 1) given that the recognition task required choosing between stimuli differing on only one feature within a brief response period. On this more difficult task, it is not surprising that people with mental retardation would have difficulty maintaining attention and performing at high levels of accuracy across a large number of test trials (cf. Hoffman et al., in press).

The findings from this study support the view that face recognition is a relatively spared visual spatial ability in adolescents and adults with WMS (Bellugi et al., 1992, 1994). They complement recent research showing that adolescents with WMS also have intact perception of biological motion, on a task which entailed spatial integration of moving light-point signals into a unitary percept of a human figure (Jordan, Reiss, Hoffman, & Landau, 2002). Despite their profound visual-spatial impairment, people with WMS are selectively spared in those components of visual information processing that involve socially significant stimuli – faces and biological motion. This selective sparing may be related to the unusually strong attention and interest in other people shown by infants and children with WMS (Jones et al., 2000; Mervis & Bertrand, 1997), which are hallmark features of this disorder.

Our study does not provide evidence on how face recognition skills develop in people with WMS, though it is unlikely that differences in the amount of time infants with WMS spend looking at faces would lead to aberrant developmental pathways (cf. Karmiloff-Smith, 1997). Recent research on WMS, as well as other neurodevelopmental disorders (cf. Tager-Flusberg, 1999), suggests that there is much less deviance in the developmental processes and neurocognitive organization in people with genetically based disorders than has been portrayed in the literature (cf. Karmiloff-Smith, 1998; Karmiloff-Smith et al., 2002). Nevertheless, the striking profiles found in WMS in cognitive sparing and impairment both across and especially within domains provide important insights on the architecture of several cognitive systems (Hoffman et al., in press; Jordan et al., 2002; Tager-Flusberg & Sullivan, 2000). Future research on the neurobiological basis of these unique profiles in visual perception and other cognitive domains in WMS and other genetic disorders has the potential to offer unique perspectives for the field of cognitive neuroscience.

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