JIDR Journal of Intellectual Disability Research
Published on behalf of mencap and in association with IASSID

Journal of Intellectual Disability Research doi: 10.1111/j.1365-2788.2011.01426.x

VOLUME 55 PART 11 pp 1034–1042 NOVEMBER 2011

# Featural versus configural face processing in a rare genetic disorder: Williams syndrome

**L. Isaac**[1,2] **& A. Lincoln**[1]

1 *Department of Clinical Psychology, California School of Professional Psychology of Alliant International University, San Diego, California, USA*
2 *Salk Institute for Biological Studies, Laboratory for Cognitive Neuroscience, La Jolla, California, USA*

## Abstract

*Background* Williams syndrome (WMS) is a rare genetic disorder with an estimated prevalence of 1 in 20 000 live births. Among other characteristics, WMS has a distinctive cognitive profile with spared face processing and language skills that contrasts with impairment in the cognitive domains of spatial cognition, problem solving and planning. It remains unclear whether individuals with WMS process faces using a featural strategy that focuses on features or a configural strategy that takes into consideration the contour of a face and spatial relations between features.
*Methods* To investigate face processing in WMS, the tasks specifically probe unfamiliar face matching by using a design that includes manipulations in face presentation (thatcherised and non-thatcherised), face orientation (upright and inverted) and face valence (happy and neutral expression) in a match-to-target face recognition design. The sample consisted of 20 participants with WMS, 10 participants with non-specific developmental delay (IQ-matched) and 10 normal control participants (chronological age-matched).
*Results* Similar to normal controls, WMS performed best when faces were presented upright. The results show while the WMS group did not perform as well as their typically developing counterparts, they did significantly better than the IQ-matched developmentally delayed group. WMS did not show an accuracy advantage for inverted faces commonly understood as an index for featural face processing, nor did they perform better on thatcherised inverted face conditions whereby featural processing is forced. Furthermore, no accuracy advantage was observed for positively valenced (happy) faces in the WMS group.
*Conclusion* These results are consistent with previous work showing a configural face processing approach in WMS, a strategy that is also utilised by normal controls.

Correspondence: Dr Linda Isaac, Department of Clinical Psychology, California School of Professional Psychology of Alliant International University, San Diego, CA 92131, USA (e-mail: lindaisaacphd@gmail.com).

**Keywords** cognitive behaviour, intellectual disability, learning disability, Williams syndrome

## Introduction

Faces are extraordinarily rich sources of information. A single glance can reveal a person's age, sex, mood and familiarity. These cues constitute crucial reference points for guiding behavioural responses. Exploring face processing strategies provides insights into the way individuals encode and recall facial identity as well as into how deviations from such strategies influence interpersonal communication. One source of such anomaly in face processing has been documented in Williams syndrome (WMS). WMS is a genetic neurodevelopmental disorder caused by the absence of approximately 28 genes on chromosome 7q11.23 (Korenberg *et al.* 2000). A salient cognitive feature of WMS is the marked impairment of visuospatial abilities as demonstrated on drawing tasks (e.g. Bellugi *et al.* 1990) contrasted with the relative sparing of face and object recognition. Specifically, persons with WMS enjoy achievement scores in the normal range on standardised face tasks (Bellugi *et al.* 1988; Udwin & Yule 1991). Despite their accuracy in face processing tasks, there is divergence in the literature as to how people with WMS attend to faces. Some have proposed that persons with WMS use a featural strategy with consideration for local features only (e.g. Karmiloff-Smith *et al.* 2004), while others have concluded that people with WMS adopt a configural strategy (Tager-Flusberg *et al.* 2003) defined by consideration for overall face contour and spatial relations between internal face features.

Until the classic paper by Yin (1969), adult expertise in face recognition was unquestionable. Yin's (1969) influential experiment of testing adult subjects' ability to recognise and identify faces presented upside down created a discrepancy in what was commonly understood as adult expertise. Yin (1969) showed that healthy subjects had poorer accuracy and longer reaction times when instructed to identify faces upside down relative to upright faces, a phenomenon termed the *inversion effect*. Configural analysis is believed to be compromised with vertically inverted faces and under these circumstances, the visual system is forced to apply a featural mode of processing. Configural processing is defined as a strategy that takes into consideration the global contour of a face as well as the spatial details between the features of a face for recognition (Rakover 2002). It is contrasted by the featural mode of face processing, which requires perception of facial features (Young *et al.* 1987; Schwarzer 2000) with an emphasis on internal or local characteristics of a face. There is a consensus in the literature that adult expertise in face processing is experience-dependent, and this expertise is developed in the context of conditions conducive to the use of a configural face processing strategy. Specifically, adults are accurate and efficient at processing human faces when the following two conditions are met: (1) when faces are presented in the upright position (a condition that is most familiar to them), and (2) when information about the contour of a face, and the spatial relations of features are made available.

There is little doubt that typically developing individuals rely on configural strategies to process faces (Bukach *et al.* 2006), and that configural strategies are disrupted with upside faces resulting in inversion effects (Tversky & Krantz 1969; Sergent 1984; Diamond & Carey 1986). These findings have led to innovative approaches of further investigation into the inversion effect. For instance, in 1993, Bartlett and Searcy used Thompson's (1980) thatcherised illusion which is created by rotating the mouth and the eyes to produce a grotesque facial expression to test the hypothesis that inversion disrupts configural processing. When presented upside down, thatcherised faces appear normal. The authors contend that this may be due to a decrease in configural and an increase in featural processing strategies. Thus, thatcherised faces provide strong evidence that configural processing is disrupted when features are inverted. The major finding of this 1980 experiment was that inversion of features (thatcherisation) impaired configural processing, forcing participants to process featural information. This study lends support for a dual-processing strategy, one mode specialised for featural information and the second for configural information. These results have been reliably replicated by others (e.g. Murray *et al.* 2000; Maurer *et al.* 2002; Boutsen & Humphreys 2003). Taken together, these findings demonstrate that even typically developing persons' ability to process faces is highly susceptible to subtle deviations from the upright position.

There continues to be disagreement in the literature about the nature of face processing abilities in persons with WMS. Specifically, the debate continues as to whether individuals with WMS process human faces using a featural/local strategy or a configural/global strategy with evidence supporting both a featural approach (Deruelle *et al.* 1999; Karmiloff-Smith *et al.* 2004) and a configural approach for faces (Tager-Flusberg *et al.* 2003; Deruelle *et al.* 2006; Rose *et al.* 2007). The present study specifically aimed to investigate if people with WMS process human faces using a featural or a configural strategy. It was expected that by manipulating the orientation, thatcherisation and emotional features of facial stimuli, the present study would shed new light on the face processing strategy utilised by individuals with WMS. In addition, given the hypersociability factor consistently reported in WMS (Jones *et al.* 2000), it was also predicted that individuals with WMS would exhibit a preference for happy faces over neutral faces and that this preference would produce an attentional bias towards processing happy faces (Dodd & Porter 2010).

The present study specifically examined the following hypotheses:

**1** Participants with WMS will show a stronger use of the configural processing strategy than the featural processing strategy, as evidenced by higher accuracy scores for upright faces compared to inverted faces.

**2** Participants with WMS will show less accuracy for faces compared to normal controls (NCs), but greater accuracy for faces compared to the IQ-matched developmentally delayed (DD) participants.

**3** Participants with WMS will perform best on conditions that are upright in orientation and happy in emotional expression.

## Method

### Participants

All WMS and DD participants were recruited from the Salk Institute for Biological Studies, Laboratory for Cognitive Neuroscience. All WMS participants were clinically diagnosed with the disorder using criteria developed by the Williams Syndrome Association Medical Advisory Board. Specifically, WMS inclusion was restricted to those participants with confirmed diagnosis via fluorescence in situ hybridisation DNA testing. Any participant who had a history of medical or neurological abnormality more severe than what is typically found in the syndrome was excluded from the present study. For instance, participants with histories of seizures or stroke were not included. All participants included in the study were English-speaking and had hearing and corrected vision, when needed. Healthy NCs were recruited from the community through newspaper and Internet advertisements. NCs were free of any present or past neurological or psychological disorder.

There were 40 participants in this study. The final sample consisted of 20 participants with WMS, 10 DD subjects and 10 NCs. Participants included 22 men (55%) and 18 women (45%) ranging in age from 18 years of age to 51 years of age (total group mean = 31.21, SD = 9.24). Peabody Picture Vocabulary Test-III (PPVT-III) scores for the total group ranged from a low of 39.00 to a high of 112.00 (M = 81.62, SD = 19.56), while Benton-Upright scores ranged from 56.00 to 119.00 (M = 89.32, SD = 17.11). Differences in the average age of each of the three groups were tested and the WMS group was found to have a significantly lower mean age than the DD group ($t = 3.341$, $P = 0.002$). The mean ages of the WMS group and the NC group did not differ significantly. The groups did not differ significantly on gender ($\chi^2 = 1.212$, $P = 0.545$). There were 12 men (60%) and eight women (40%) in the WMS group, six men (60%) and four women (40%) in the DD group, and four men (40%) and six women (60%) in the NC group.

### Assessment measures

The assessment measures for this study included the PPVT-III, and the Benton-Upright Faces Task. In order to assess for receptive vocabulary and face processing ability, we administered both the PPVT-III and the Benton Face Task measures. Importantly, the profile of WMS includes verbal memory impairment, thus the PPVT-III is a preferred measure for estimating language functioning in WMS because it is free of word retrieval demands. Healthy NCs who obtained a PPVT-III standard score above 115 were excluded from this study.

### Materials, apparatus and instructions

Thatcherised faces were created by inverting the mouth and both eyes with Adobe Photoshop 2007 software system. Adobe software was also used to place all face stimuli in an oval frame to entirely eliminate hair and background information. All face stimuli were retrieved from a face database at the Salk Institute for Biological Studies, Laboratory for

Cognitive Neuroscience. Stimuli were presented in digitised format on a computerised stimuli program (PsyScope) to standardise display time, response time, response style and proximity of faces as they were presented. Participants were seated in front of a computer at a distance of 80 cm from the computer screen. In randomised order, participants were presented with 24 trials per stimulus condition consisting of all possible eight experimental conditions (Table 1).

Participants were presented with a target face in the centre, upper part of the display for 200 ms and were asked to look carefully at each face that was presented. Below this image, two response choice faces of the same individual appeared until the participant responded (no time limit was imposed). Participants were instructed to identify which of the two faces exactly matched the target face by pointing to the face. The experimenter was responsible for pressing one of the two labelled keys on a keyboard (the 'Z' button to select the left face response choice or the '/' button to select the right face response choice). In half of the trials, all faces were upright, and in the other half, they were inverted. The side of the correct response choice was counterbalanced. All participants underwent a training procedure with non-human face stimuli (animals), to ensure they understood the test format and appropriate use of the response buttons. The training stimuli closely resembled the final test stimuli (e.g. upright and inverted cat faces). Feedback was given during the training phase to ensure that all participants understood the match-to-target procedure.

**Table 1** Eight experimental conditions

| Affect | Presentation | Orientation |
|---|---|---|
| Happy | Thatcherised | Upright |
| Neutral | Thatcherised | Upright |
| Happy | Non-thatcherised | Upright |
| Happy | Thatcherised | Inverted |
| Neutral | Non-thatcherised | Upright |
| Neutral | Thatcherised | Inverted |
| Happy | Non-thatcherised | Inverted |
| Neutral | Non-thatcherised | Inverted |

### Target presentation and exposure time

Exposing target stimuli at 200 ms is an optimal time for differentiating a featural versus a configural bias for face processing. Studies which have not imposed a time limit for stimuli presentation have been critiqued by others for elongating exposure and thus forcing a configural processing strategy (Karmiloff-Smith 1997), while others have not implemented enough exposure time (e.g. 150 ms) to the target stimuli, consequently forcing a featural mode of face processing (Tager-Flusberg *et al.* 2003).

### Data analysis

The comparison between the groups for differences on measures of overall facial perception accuracy (Benton-Upright Face Task) and receptive vocabulary (PPVT-III) was conducted by means of a one-way analysis of variance (ANOVA) with group (WMS, DD, NC) as the factor. Hypothesis 1, comparing the accuracy scores of the WMS group for upright faces versus inverted faces, was tested by means of a within-group ANOVA with face orientation as the factor. Hypothesis 2, comparing the accuracy score of the WMS group to those of the DD and NC groups, was tested by conducting an ANOVA of overall accuracy scores with group as the factor. Hypothesis 3, comparing the accuracy scores of WMS group for happy versus neutral facial expression in both upright and inverted orientation, was tested by means of a within-group ANOVA, using the two facial expressions (happy, neutral) and orientation (upright, inverted) as levels of the within-group ANOVA. *Post hoc* comparisons were conducted after an initial finding of a significant overall *F*-value.

## Results

### Descriptive cognitive data

*Benton-Upright Face Task*

On the Benton-Upright Face Task, the overall ANOVA result for group (WMS, DD and NC) was significant ($F_{2,35} = 10.35$, $P = < 0.001$). The Tukey *post hoc* comparison test found the mean for the WMS group to be significantly ($P < 0.001$) higher than that for the DD group. No other pairwise

**Table 2** Means and standard deviations for age, PPVT and Benton-U

| Group | *n* | Age | | PPVT | | Benton-U | |
|---|---|---|---|---|---|---|---|
| | | Mean | SD | Mean | SD | Mean | SD |
| Williams syndrome | 20 | 27.57 | 5.18 | 78.95 | 11.63 | 94.70 | 14.53 |
| Developmentally delayed | 10 | 36.40 | 9.40 | 65.20 | 21.30 | 71.60 | 13.65 |
| Normal (controls) | 10 | 33.30 | 12.65 | 103.40 | 8.75 | 96.30 | 12.99 |
| Total | 40 | 31.21 | 9.24 | 81.62 | 19.56 | 89.32 | 17.11 |

PPVT, Peabody Picture Vocabulary Test; Benton-U, Benton-Upright.

group differences were found to be significant on this measure of facial perception abilities.

### *Peabody Picture Vocabulary Test-III*

The overall ANOVA result for group on the PPVT-III of receptive vocabulary was statistically significant ($F_{2,35} = 20.27$, $P = < 0.001$). The Tukey *post hoc* test found the difference between the WMS and DD groups to be non-significant ($P = 0.111$). However, the same test found that the NC group scored significantly higher than both the WMS group and the DD group ($P < 0.001$ in both cases; Table 2).

## Orientation, thatcherisation and emotional expression

In order to ascertain to what degree orientation, thatcherisation and emotional expression of face stimuli influenced performance (accuracy), paired *t*-tests were computed between accuracy scores under orientation, thatcherisation and emotional expression both separately and in combination. These comparisons were conducted for the sample as a whole as the hypotheses addressed these same effects within all three groups. The results of these comparisons indicate that accuracy scores were significantly influenced by orientation ($t_{1,39} = 8.98$, $P = < 0.001$). Accuracy scores for all three groups were lower in the inverted orientation (M = 59.22, SD = 17.6) compared to the upright orientation (M = 86.96, SD = 14.6). Strikingly, the performance of NCs was also significantly impacted ($P = 0.040$) by the inversion effect on this task (upright: M = 98.15, SD = 3.1; inverted: M = 71.60, SD = 10.7). Thatcherisation also had a significant impact on performance ($t_{1,39} = 2.12$, $P = 0.04$), with higher accuracy for all three groups when faces were both thatcherised and upright. However, thatcherisation did not impact performance when face stimuli were both thatcherised and inverted ($P = 0.284$). Facial emotional expression (happy, neutral) did not have an impact on accuracy performance ($P = 0.349$) across the groups.

In summary, examination of the variables orientation, thatcherisation and emotional expression indicates the main source of the differences is attributable to face orientation (i.e. inversion). Thatcherisation added at most, a slight increment to the observed differences. Specifically, the influence of thatcherisation was highly dependent on the variable orientation. Finally, emotional expression did not impact the results.

## Tests of hypotheses

Hypothesis 1 predicts that participants with WMS will exhibit higher accuracy scores for upright faces compared to inverted faces, which would be indicative of a configural face processing strategy in this group. This hypothesis was tested by a within-group ANOVA. The results of this ANOVA indicate that differences between the orientations of the facial stimuli in the WMS group were highly significant ($F_{1,19} = 57.73$, $P = < 0.001$, $\eta^2 = 0.75$). As predicted, mean accuracy was significantly higher for the upright facial stimuli (M = 88.54, SD = 13.6) than for the inverted facial stimuli (M = 56.11, SD = 13.9). Also in line with our prediction, the within-group analysis for thatcherisation and non-thatcherisation revealed that persons with WMS performed equally in inverted thatcherised and inverted non-thatcherised face conditions ($P$-values $\geq 0.05$). In other words, no advantage in

**Table 3** Group accuracy for eight experimental conditions

| | Group | | | | | |
|---|---|---|---|---|---|---|
| | Williams syndrome | | Developmentally delayed | | Normal controls | |
| Experimental condition | Mean | Standard deviation | Mean | Standard deviation | Mean | Standard deviation |
| Happy/thatch/upright | 92.40 | 15.92 | 90.00 | 21.08 | 96.60 | 10.75 |
| Neutral/thatch/upright | 87.85 | 18.11 | 64.90 | 9.30 | 98.00 | 6.32 |
| Happy/thatch/inverted | 49.80 | 15.90 | 56.50 | 24.73 | 82.90 | 24.99 |
| Neutral/thatch/inverted | 61.55 | 31.11 | 51.50 | 9.30 | 70.00 | 34.96 |
| Happy/non-th/upright | 89.05 | 20.60 | 69.10 | 28.35 | 100.00 | 0.00 |
| Happy/non-th/inverted | 47.40 | 16.39 | 50.30 | 17.04 | 66.90 | 24.51 |
| Neutral/non-th/upright | 84.85 | 20.32 | 66.40 | 15.22 | 98.00 | 6.32 |
| Neutral/non-th/inverted | 65.70 | 29.39 | 54.00 | 16.47 | 66.60 | 33.33 |

Emotion, happy or neutral; thatch, thatcherised; non-th, non-thatcherised; orientation, upright or inverted.

accuracy performance was observed in the condition which forces featural processing of faces (i.e. inverted thatcherised).

Hypothesis 2 predicts that people with WMS will show less accuracy for faces compared to the NC group, but greater accuracy for faces compared to the IQ-matched DD group. This was tested by conducting an ANOVA of overall accuracy scores with group as the factor. *Post hoc* tests were conducted between the pairs of groups specified in the hypothesis. The overall *F*-test for the group effect was highly significant ($F_{2,37} = 11.07$, $P = < 0.001$, $\eta^2 = 0.37$). As expected, the *post hoc* comparison of accuracy means between the WMS group and the NC group was significant ($P = 0.012$), with overall accuracy of the NC group being higher (M = 84.88, SD = 12.48) than that of the WMS group (M = 72.33, SD = 9.97). Consistent with our prediction, the *post hoc* tests also revealed that the overall accuracy of the WMS group, compared to their IQ-matched counterparts (DD group), was significant ($P = 0.051$) with higher overall accuracy performance for the WMS group compared to the DD group (M = 62.84, SD = 9.47).

Hypothesis 3 predicted that people with WMS would perform best on conditions that are upright in orientation and happy in emotional expression. The test of this hypothesis consisted of a within-group ANOVA, using the two facial emotional expressions (happy, neutral) and orientation (upright, inverted) as levels of the within-group factors. The results of this analysis indicate that significant differences do not exist on the basis of facial emotion expression ($P = > 0.05$). Contrary to our prediction, happy facial expressions did not improve overall accuracy for the WMS group (Table 3).

## Discussion

We aimed to examine whether individuals with this rare genetic condition process faces using a configural strategy utilised by adults without disorders or if they deviate from this process using an alternative mode: featural face processing. Consideration for the impact of facial valence on processing strategy was also incorporated in this study in light of the well-documented social approach tendencies in WMS (Bellugi *et al.* 1999; Jones *et al.* 2000; Doyle *et al.* 2004; Järvinen-Pasley *et al.* 2010).

Performance of persons with WMS was compared to those with a non-specific developmental delay group matched by mental age and typically developing individuals matched by chronological age. The experiments we have described allow us to make some interesting inferences about face processing in this unique genetic disorder. Our results help to reconcile an important inconsistency between past experimental findings with respect to face processing in WMS. Specifically, findings from the present study suggest that the face processing strategy employed by persons with WMS is configural. Their processing strategy parallels that of persons without intellectual constraints showing a clear departure from previous research that points to a deviant, featural face processing strategy in this genetic disorder (e.g. Bihrle *et al.* 1989; Karmiloff-Smith 1997; Karmiloff-Smith *et al.* 2004).

Comparisons indicated all of the groups had higher scores for upright face presentations and all groups, including NCs, showed a significant drop in accuracy scores for inverted faces. This is in agreement with many reports showing inversion effects for all groups (Bartlett & Searcy 1993; Farah *et al.* 1995; Freire *et al.* 2000; Murray *et al.* 2000; Barton *et al.* 2003; Itier & Taylor 2004). Our results clearly align with those who reject the hypothesis that face processing is atypical in WMS (e.g. Tager-Flusberg *et al.* 2003). The WMS group did not differ from the NC group on conditions differing in presentation only (thatcherised and non-thatcherised). The present thatcherisation results are inconsistent with the most recent explorations of face processing in WMS using the thatcher illusion (Riby *et al.* 2009). The present results show the WMS group did not show an advantage in processing inverted thatcherised faces (an index for featural face processing) offering further evidence of a configural face processing strategy in this group (e.g. Bellugi *et al.* 2000). In other words, individuals with WMS use a face processing strategy that is analogous to typically developing people despite their intellectual compromises challenging the assumption that this group has an abnormal approach to processing human faces. Importantly, several aspects of the results suggest WMS participants were less accurate than NCs, signifying they found the experimental task more difficult than NCs, but less challenging than the IQ-matched DD group. Thus, it may be extrapolated that face processing in WMS is also experience-dependent, similar to normal populations (Maurer *et al.* 2002). Because faces are usually seen upright, it is not surprising orientation was a critical variable for a face processing system developed over many years of experience. Similar to typically developing adults who rely on upright, configural information to recognise a face, this relative face processing strength in WMS is equally related to their experience and high sensitivity for human faces (Schwaninger *et al.* 2003). Previous research has attempted to explain why faces are special to those with WMS by referring to the hypersociability feature of this syndrome (Pober & Dykens 1996; Bellugi *et al.* 1999; Doyle *et al.* 2004). The indiscriminate social approach tendencies observed in WMS lead us to suggest that faces are special stimuli and the social relevance that faces contain may help to explain this so-called relative strength in WMS. An explanation for this finding can perhaps best be described by turning to the neuroimaging dialogue. Recently, a study by Golarai *et al.* (2010) found the fusiform face area, implicated in face recognition, is relatively enlarged in the WMS brain. This neuroanatomical finding can help explain face recognition proficiency among those with WMS.

In the present study, WMS did not show an advantage in performance accuracy for happy faces compared to neutral faces and as such no significant differences were found for this dimension of the study. One explanation for this result comes from evidence that approach behaviour in WMS is not restricted to positive (happy) facial valence as it is in typically developing people (Martens *et al.* 2009; Paul *et al.* 2009).

In conclusion, it remains undisputed that face processing is a relative strength for persons with WMS. This is especially evidenced in the finding that WMS performed significantly better than the IQ-matched group (DD). The unique cognitive disparities in WMS have inspired research on how the relative strength of face processing ability mediates or compensates for the relative weaknesses (e.g. visual–spatial processing). On the basis of our findings, people with WMS process faces using a configural strategy, similar to typically developing people.

## Limitations

Several limitations of our study merit comment. The first limitation of the study is a small sample size. Second, reaction times were not collected, thus a speed accuracy trade-off cannot be fully determined. Lastly, our design was also limited by the presentation of static faces with a single viewing perspective. We recommend a follow-up study that better simulates the demands of everyday face processing tasks such as the inclusion of a wider face-valence range (e.g. angry and surprised emotional faces) and variations in gaze direction (direct and averted). The pragmatic significance of such understanding lies in helping persons with WMS better differentiate between familiar and non-familiar persons and emotional expressions that serve to better facilitate approach-avoidance response in social contexts common to real settings and critical to guiding adaptive behaviour. For instance, one can quickly fathom why an angry face with a direct gaze bears greater personal relevance to an observer than an angry facial expression with an averted gaze.

## References

Bartlett J. C. & Searcy J. (1993) Inversion and configuration of faces. *Cognitive Psychology* **25,** 281–316.

Barton J. J., Deepak S. & Malik N. (2003) Attending to faces: change detection, familiarization, and inversion effects. *Perception* **32,** 15–28.

Bellugi U., Wang P. P. & Jernigan T. L. (1988) Williams syndrome: an unusual neuropsychological profile. In: *Atypical Cognitive Deficits in Developmental Disorders: Implications for Brain Function* (eds S. Broman & J. Grafman). Lawrence Erlbaum Associates, Hillsdale, NJ.

Bellugi U., Bihrle A., Jernigan T., Trauner D. & Doherty S. (1990) Neuropsychological, neurological, and neuroanatomical profile of Williams syndrome. *American Journal of Medical Genetics. Supplement* **6,** 115–25.

Bellugi U., Adolphs R., Cassady C. & Chiles M. (1999) Towards a neural basis for hypersociability in a genetic syndrome. *Neuroreport* **10,** 1–5.

Bellugi U., Lichtenberger L., Jones W., Lai Z. & St George M. (2000) The neurocognitive profile of Williams Syndrome: a complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience* **12,** 7–29.

Bihrle A. M., Bellugi U., Delis D. & Marks S. (1989) Seeing either the forest or the trees: dissociation in visuospatial processing. *Brain and Cognition* **11,** 37–49.

Boutsen L. & Humphreys G. W. (2003) The effect of inversion on the encoding of normal and 'thatcherized' faces. *The Quarterly Journal of Experimental Psychology* **56,** 955–75.

Bukach C. M., Bub D. N., Gauthier I. & Tarr M. J. (2006) Perceptual expertise effects are not all or none: spatially limited perceptual expertise for faces in a case of prosopagnosia. *Journal of Cognitive Neuroscience* **18,** 48–63.

Deruelle C., Mancini J., Livet M. O., Cassé-Perrot C. & de Schonen S. (1999) Configural and local processing of faces in children with Williams syndrome. *Brain and Cognition* **41,** 276–98.

Deruelle C., Rondan C., Mancini J. & Livet M. O. (2006) Do children with Williams syndrome fail to process visual configural information? *Research in Developmental Disabilities* **27,** 243–53.

Diamond R. & Carey S. (1986) Why faces are and are not special: an effect of expertise. *Journal of Experimental Psychology. General* **115,** 107–17.

Dodd H. F. & Porter M. A. (2010) I see happy people: attention bias towards happy but not angry facial expressions in Williams syndrome. *Cognitive Neuropsychiatry* **21,** 1–19.

Doyle T. F., Bellugi U., Korenberg J. R. & Graham J. (2004) 'Everybody in the world is my friend'. Hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics* **124,** 263–73.

Farah M. J., Tanaka J. W. & Drain H. M. (1995) What causes the face inversion effect? *Journal of Experimental Psychology. Human Perception and Performance* **21,** 628–34.

Freire A., Lee K. & Symons L. (2000) The face-inversion effect as a deficit in the encoding of configural information: direct evidence. *Perception* **29,** 159–70.

Golarai G., Hong S., Haas B. W., Galaburda A. M., Mills D. L., Bellugi U. *et al.* (2010) The fusiform face area is enlarged in Williams syndrome. *Journal of Neuroscience* **30,** 6700–12.

Itier R. J. & Taylor M. J. (2004) Face inversion and contrast-reversal effects across development: in contrast to the expertise theory. *Developmental Science* **7,** 246–60.

Järvinen-Pasley A., Adolphs R., Yam A., Hill K. J., Grichanik M., Reilly J. *et al.* (2010) Affiliative behavior in Williams syndrome: social perception and real-life social behavior. *Neuropsychologia* **48,** 2110–19.

Jones W., Bellugi U., Lai Z., Chiles M., Reilly J., Lincoln A. *et al.* (2000) Hypersociability in Williams Syndrome. *Journal of Cognitive Neuroscience* **12,** 30–46.

Karmiloff-Smith A. (1997) Crucial differences between developmental cognitive neuroscience and adult neuropsychology. *Developmental Neuropsychology* **13,** 513–24.

Karmiloff-Smith A., Thomas M., Annaz D., Humphreys K., Ewing S., Brace N. *et al.* (2004) Exploring the Williams syndrome face-processing debate: the importance of building developmental trajectories. *Journal of Child Psychology and Psychiatry, and Allied Disciplines* **45,** 1258–74.

Korenberg J. R., Chen X. N., Hirota H., Lai Z., Bellugi U., Burian D. *et al.* (2000) VI. Genome structure and cognitive map of Williams syndrome. *Journal of Cognitive Neuroscience* **12,** 89–107.

Martens M. A., Wilson S. J., Dudgeon P. & Reutens D. C. (2009) Approachability and the amygdala: insights from Williams syndrome. *Neuropsychologia* **47,** 2446–53.

Maurer D., Le Grand R. & Mondloch C. J. (2002) The many faces of configural processing. *Trends in Cognitive Sciences* **6,** 255–60.

Murray J. E., Yong E. & Rhodes G. (2000) Revisiting the perception of upside-down faces. *Psychological Science* **11,** 492–6.

Paul B. M., Snyder A. Z., Haist F., Raichle M. E., Bellugi U. & Stiles J. (2009) Amygdala response to faces parallels social behavior in Williams syndrome. *Social Cognitive and Affective Neuroscience* **4,** 278–85.

Pober B. R. & Dykens E. M. (1996) Williams syndrome: an overview of medical, cognitive and behavioral features. *Child and Adolescent Psychiatric Clinics of North America* **5,** 929–43.

Rakover S. (2002) Featural vs. configurational information in faces: a conceptual and empirical analysis. *British Journal of Psychology* **93,** 1–30.

Riby D. M., Riby L. M. & Reay J. L. (2009) Differential sensitivity to rotations of facial features in the Thatcher illusion. *Psychological Reports* **105**(Pt 1), 721–6.

Rose F. E., Lincoln A. J., Lai Z., Ene M., Searcy Y. M. & Bellugi U. (2007) Orientation and affective expression effects on face recognition in Williams syndrome and autism. *Journal of Autism and Developmental Disorders* **37,** 513–22.

Schwaninger A., Carbon C. C. & Leder H. (2003) Expert face processing: specialization and constraints. In: *Development of Face Processing* (eds G. Schwarzer & H. Leder), pp. 81–97. Hogrefe, Göttingen.

Schwarzer G. (2000) Development of face processing: the effect of face inversion. *Child Development* **71,** 391–401.

Sergent J. (1984) An investigation into component and configural processes underlying face perception. *British Journal of Psychology* **75,** 221–42.

Tager-Flusberg H., Plesa-Skwerer D., Faja S. & Joseph R. M. (2003) People with Williams syndrome process faces holistically. *Cognition* **89,** 11–24.

Thompson P. (1980) Margaret Thatcher: a new illusion. *Perception* **9,** 483–4.

Tversky A. & Krantz D. H. (1969) Similarity of schematic faces: a test of interdimensional additivity. *Perception and Psychophysics* **5,** 124–8.

Udwin O. & Yule W. (1991) A cognitive and behavioral phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology* **13,** 232–44.

Yin R. K. (1969) Looking at upside-down faces. *Journal of Experimental Psychology* **81,** 141–5.

Young A. W., Hellawell D. & Hay D. C. (1987) Configurational information in face perception. *Perception* **16,** 747–59.

*Accepted 1 April 2011*