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Toddlers with Williams Syndrome Process Upright but Not Inverted Faces Holistically

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Abstract

Holistic processing of upright, but not inverted, faces is a marker of perceptual expertise for faces. This pattern is shown by typically developing (TD) individuals beginning at age 7 months. Williams syndrome (WS) is a rare neurogenetic developmental disorder characterized by extreme interest in faces from a very young age. Research on the effects of inversion on holistic processing of faces by older children and adults with WS has produced mixed results. Younger children with WS were not included in these previous studies. Using the habituation switch paradigm, we demonstrated that 15 – 35-month-olds with WS process upright, but not inverted, faces holistically. This study provides evidence of perceptual expertise for faces in individuals with WS early in life.

Keywords

face perception; holistic processing; inversion effect; perceptual expertise; Williams syndrome; developmental disability

Williams syndrome (WS) is a neurodevelopmental disorder caused by a deletion of ~25 genes on chromosome 7q11.23 (Hillier et al., 2003). Young children with WS have developmental delay; older children typically have intellectual disability or learning difficulties. Individuals with WS are extremely interested in people, faces, and face-to-face interactions and attend to faces more than do typically developing (TD) children or adults (e.g., Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Mervis et al., 2003; Riby & Hancock, 2009b). Results of recent eye-tracking studies demonstrate that when shown pictures of social scenes, school-aged children and adults with WS spend more time fixated on human faces than do either TD individuals matched for chronological age (CA) or TD children matched for nonverbal reasoning ability (mean CA 7.5 years) (Riby & Hancock, 2008). In contrast, individuals with autism shown the same pictures spend significantly less time attending to faces than do TD individuals matched for CA or nonverbal reasoning ability (Riby & Hancock, 2008). Similar findings were obtained when the stimuli were pictures of natural scenes (Riby & Hancock, 2009a) or videos (Riby & Hancock, 2009b).

This pattern of strong interest in faces emerges early in life for individuals with WS. Mervis et al. (2003) found that compared to TD infants and toddlers matched for either CA or developmental age, infants and toddlers with WS focused longer and more intensely on the face of the adult with whom they were playing (see also Jones et al., 2000). Furthermore, when being examined by a physician, 8- to 35-month-olds with WS—unlike TD children of the same CA or children with other genetic syndromes of the same CA—continued to focus

intently on the adult's face even when the adult shifted attention from the child's face to another target (Mervis et al., 2003). While there is agreement that people with WS have a heightened interest in faces beginning in infancy (e.g., Mervis et al., 2003) and that hypersociability is commonly demonstrated beginning in the toddler period (e.g., Doyle et al., 2004), there are no published studies addressing the face-processing mechanisms of children with WS younger than 4 years old. In the present study, we investigated the face processing abilities of toddlers with WS. In the remainder of the introduction we summarize prior findings of expert-like face processing by both adults and infants in the general population. We then consider findings from similar studies of individuals with WS and outline the present study.

The “face inversion effect” is a well-known phenomenon that describes the fact that adults in the general population are better at recognizing, remembering, and discriminating faces in the upright, canonical orientation compared to inverted faces (e.g., Valentine, 1988; Rossion & Gauthier, 2002). This inversion effect is considered a sign of perceptual expertise resulting from the use of different modes of processing when viewing faces in the upright orientation than when viewing the same faces in the inverted orientation (e.g., Farah, Tanaka, & Drain, 1995; Freire, Lee, & Symons, 2000; see also Rossion, 2008, 2009; Rossion & Gauthier, 2002). In particular, individuals in the general population have been shown to use holistic processing (i.e., processing the combination of facial features) and 2nd-order configural processing (i.e., processing the spacing between facial features) when viewing upright faces but to use featural, or piecemeal, processing when viewing inverted faces (for review, see Maurer, LeGrand, & Mondloch, 2002; see also Rossion, 2008, 2009). Developmentally, there is evidence that such effects begin during infancy. For example, previous research with TD infants indicates that inversion begins to affect holistic face processing during the first year of life. Using the visual habituation “switch” paradigm, evidence was found that TD 3-month-olds processed both upright and inverted faces featurally, and TD 4-month-olds processed both upright and inverted faces holistically (Cashon & Cohen, 2004). TD 7–8-month-olds, however, showed the expert-like pattern of processing upright faces holistically but inverted faces featurally (Cohen & Cashon, 2001; Ferguson, Kulkofsky, Cashon, & Casasola, 2009). Hereinafter, we refer to this pattern as the “expert-like pattern of holistic face processing.”

Although there have been a number of studies on face processing by individuals with WS (e.g., Karmiloff-Smith et al., 2004; Martens, Wilson, & Reutens, 2008; Riby & Porter, 2010; Rose et al., 2007), only two studies have addressed the question of whether individuals with WS show the expert-like pattern of holistic face processing (Tager-Flusberg, Plesa Skwerer, Faja, & Joseph, 2003; Annaz, Karmiloff-Smith, Johnson, & Thomas, 2009). To address this question, Tager-Flusberg et al. (2003) tested adolescents and adults with WS using the part-whole task (Tanaka & Farah, 2003) commonly used to study face processing by adults in the general population. They found that individuals with WS showed the expert-like pattern of holistic face processing exhibited by individuals in the general population: upright faces were processed holistically but inverted faces were processed featurally. Annaz et al. (2009) used a slightly different procedure to study the face processing abilities of a group of children with WS aged 4 – 12 years old. In contrast to Tager-Flusberg et al., Annaz et al. did not find a statistically significant difference in performance between the upright and inverted conditions. On that basis, they concluded that children with WS have a bias toward processing both upright and inverted faces featurally. Although one interpretation of the disparate findings between the two studies is that the expert-like pattern of holistic face processing develops between childhood and adolescence in individuals with WS, another possibility is that the difference in results is due to differences between the tasks that were used. In particular, whereas the standard part-whole paradigm tests face processing by first training participants to identify correctly a set of training faces and then replacing the

training set with the test stimuli, Annaz et al. showed the target face and the test stimuli simultaneously. Therefore, the memory component of the part-whole paradigm was eliminated. Recent studies by Richler, Cheung, and Gauthier (2011) provide evidence that the simultaneous presentation of target and test faces induces an overreliance on featural face processing strategies. Thus, it is unclear if children with WS have the expert-like pattern of face processing or even whether they are able to process upright faces holistically at all.

In the present study, we examined the ability of toddlers with WS to process upright and inverted faces. We reasoned that children with WS, who are especially interested in faces and therefore have extensive experience with them, should show the expert-like pattern of holistic face processing of upright faces but featural processing of inverted faces at a young age, even though they are developmentally delayed. Due to the rarity of this syndrome, we used a within-subjects design to test this hypothesis. Thus we compared processing of upright and inverted faces by the same group of 15 – 35-month-olds with WS. We predicted that toddlers with WS would evidence holistic processing for upright, but not inverted, faces given that: 1) TD infants evidence the expert-like holistic face processing pattern at 7 months, 2) beginning in infancy individuals with WS exhibit heightened attention to faces and therefore have heightened exposure to upright faces, and 3) perceptual expertise for faces is thought to be the result of experience (e.g., Gauthier & Nelson, 2001). We used the habituation “switch” task previously used to test TD infants (e.g., Cohen & Cashon, 2001). The switch task is based on looking times and therefore is a useful method for testing nonverbal participants.

In the habituation “switch” task, participants are first habituated to two faces, that is, they are repeatedly shown these two faces until their looking time drops significantly, indicating that these faces have become familiar to them. During the subsequent test phase, participants are shown three faces one at a time in counterbalanced order: a familiar face (one of the two faces shown during the habituation phase), a switch face (a face that is composed of the internal features of one of the habituation faces and the external features of the other; hence the internal and external features have been switched), and a novel face. The critical comparison in this design is how long participants look at the switch test face relative to the familiar test face. If participants notice the novel combination of familiar features in the switch test face, they would be expected to look longer at that face than at the familiar face. This looking pattern provides evidence of holistic processing. In the context of this task, holistic processing is operationally defined as processing the correlation between the internal and external features of a face (Cohen & Cashon, 2001).

In contrast, if participants notice that the features in the switch test face are familiar but do not notice that they have been combined in a novel way, they would be expected to treat the switch test face as familiar and therefore not demonstrate increased looking time toward it. The latter looking pattern would provide evidence of featural, or piecemeal, processing. Longer looking time at the novel face compared to the familiar face indicates that participants can distinguish between a familiar face and a completely novel face and, importantly, that they are not fatigued. [In similar studies of TD infants, longer looking time to a novel face than to a familiar face is evidenced by age 3 months (Cashon & Cohen, 2004).] Based on our hypothesis, we expected that toddlers with WS would look significantly longer at the switch face than the familiar face in the upright condition but not in the inverted condition. Based on previous results in this task with TD infants, we predicted that participants would show longer looking at the novel test face relative to the familiar test face in both orientations. This would indicate that they could detect the novel face as novel and were not fatigued.

Method

Participants

Fourteen toddlers (7 girls, 7 boys) with a genetically confirmed diagnosis of WS and normal or corrected-to-normal vision participated in this study. All participants had classic-length deletions as determined by FISH or qPCR. The racial/ethnic background of the participants was: 13 White/Non-Hispanic and 1 biracial Non-Hispanic (African American/White). Mean chronological age, after adjusting for two premature participants, was 25.7 months ($SD = 6.76$, range: 15.4 – 34.9 months).

All 14 participants successfully completed testing in both the upright condition and the inverted condition either the same day or on consecutive days. Order of presentation of the conditions was counterbalanced, as described below. Six additional babies were excluded due to: fussiness ($n = 3$, upright condition); failure to meet the habituation criterion ($n = 1$, inverted condition); or looking for the maximum trial length or more than 2 standard deviations above the mean during the familiar test trial, indicating that full habituation did not occur ($n = 1$, upright condition; $n = 1$, inverted condition).

Materials

Developmental assessment—The Mullen Scales of Early Learning (MSEL; Mullen, 1995) provides a standardized comprehensive measure of young children's abilities in the cognitive and language domains. The Early Learning Composite [ELC; similar to Developmental Quotient (DQ)] is based on performance on four scales: Visual Reception (measuring primarily nonverbal reasoning), Fine Motor (measuring primarily eye-hand coordination and visuospatial construction), Receptive Language, and Expressive Language. For the general population, the mean ELC is 100, with a standard deviation of 15 and a range from 49 – 155.

Thirteen of the 14 participants completed the MSEL. Seven participants completed the MSEL within 1 day of the day they were tested in this study. The remaining 6 participants completed the MSEL at a different time, ranging from 6 months prior to 2 months after being tested in this study (mean difference: 1.4 months). ELCs ranged from 49 (moderate developmental delay) to 82 (low average for the general population), with a mean in the mild developmental delay range ($M = 60.92$, $SD = 12.49$). This mean ELC is almost identical to the mean ELC (61.45) reported by Mervis and John (2010) for a sample of 144 toddlers and young preschoolers with WS. Thus, the intellectual functioning of the participants in the present study relative to individuals their age in the general population is consistent with that expected for very young children with WS.

Face stimuli—The complete set of stimuli included 12 color photographs of faces of college-age Caucasian females (mean size 19.3 cm \times 27.4 cm, mean visual angle of 9.4° \times 13.7°) presented on a gray background. To confirm that all faces showed neutral expressions, 20 adults in the general population rated the expression on each face on a 9-point Likert scale (1 = very sad, 3 = sad, 5 = neutral, 7 = happy, 9 = very happy). The mean rating for each face ranged from 4.9 to 5.7 with an overall mean of 5.1, indicating that all of the facial expressions were neutral. Example stimuli are shown in Figure 1.

The 12 faces included six original faces and six modified (“switched”) faces. Eight of the 12 faces were used in a previous study with TD infants (Cashon, Ha, Allen, & Barna, in press). The other four were created for this study. All stimuli were created using the methods described by Cohen and Cashon (2001) as follows: Photographs were edited in Adobe® Photoshop® to remove any jewelry present, to normalize size and brightness, and to place

the faces on a gray background. The six original faces were divided into three pairs. Switch faces were created by digitally copying the internal features from one face in a pair and pasting them onto the other face in the pair. To do this, a digital cut was made around the area above the eyes but below the eyebrows, across the cheeks, along the outside of the nose, and around the bottom of the mouth. This area was then pasted onto the other face and blended so that the new switch face appeared natural. Both original and switched faces were used as habituation and test stimuli across participants. Although all 12 face stimuli were used in both the upright and inverted conditions, no toddler saw the same set of faces in both orientations.

Procedure

Each participant was seated on an adult's lap approximately 120 cm from a Panasonic 50 color plasma screen (1024 × 576 pixels) in a dimly lit room. The adult (almost always an undergraduate assistant from another laboratory) was instructed not to interact with the toddler during testing to minimize distractions and eliminate any possible influence on the participant's direction of gaze. The adult was not aware of the research hypotheses. It is standard in our laboratory that if an adult is found to distract or influence the child during testing, that child's data are excluded. No data had to be omitted for these reasons in this study. To determine the toddler's direction of gaze, a trained researcher, who was in an adjacent room, viewed the toddler's eyes and corneal reflections on a 15" JVC color monitor connected to a Canon VC-C50i camera hidden below the plasma screen. Habit X software (Cohen, Atkinson, & Chaput, 2004), running on a Macintosh Power Mac G5 computer, was used to control stimulus presentation and calculate looking times based on the experimenter's key presses as described below. The experimenter could not see the stimuli presented to the participant during testing. All sessions were recorded to DVD. Data from all participants were tested for inter-rater reliability on the first four habituation trials, the last four habituation trials, and the three test trials. To compute inter-rater reliability, a second trained experimenter ran Habit X while viewing the DVD of each testing session. If there was a difference greater than 1 second on any of the test trials, that participant was excluded. No participant was excluded for this reason. The correlation between the looking times recorded by the live experimenter and the offline experimenter was $r = .99$.

Prior to each trial, a video of an expanding and contracting green ball accompanied by a "ding" sound played on a black background in the center of the monitor to attract the participant's attention to the center of the screen. When the baby looked at the center of the screen, the experimenter pressed the Enter key. This resulted in the attention-getter being replaced with a face stimulus, which marked the beginning of the trial. Throughout the trial, the experimenter held down the "5" key if the toddler was looking at the face; whenever the toddler was not looking at the face, the experimenter released the "5" key. Each trial lasted until the participant looked away from the screen for at least 1 continuous second or until 20 seconds of looking time had elapsed. When the trial ended, the attention-getter reappeared on the monitor. This sequence was repeated throughout the session.

During the habituation phase, each participant viewed two faces (e.g., Faces A & B) sequentially in a quasi-randomized order. The habituation phase continued until Habit X had calculated that the toddler's looking time had reached the habituation criterion, which was that the mean looking time on a sliding window of four consecutive trials was less than 50% of the mean looking time on the first four trials.

During the test phase, each participant viewed three test faces: familiar (e.g., Face A), switch (e.g., internal features of Face B combined with external features of Face A), and novel. The order of presentation of test faces was counterbalanced across participants. A within-subject design was used; participants were tested in both the upright and inverted face conditions

within a two-day period. The order of the orientation conditions was randomized by a flip of a coin. Seven participants were tested in the upright condition first; the remaining seven were tested first in the inverted condition. Each participant saw a different set of faces in each orientation.

Results

Habituation Phase

To determine if there were differences in performance during the habituation phase as a function of orientation condition, three Wilcoxon tests were conducted. The median number of trials to meet the habituation criterion was 8 in the upright condition and 10 in the inverted condition. This difference was not significant, $Z = -0.24$, $p = .806$. Additionally, we found no significant difference in average looking time on the first four habituation trials between the upright ($Mdn = 11.7$) and inverted conditions ($Mdn = 10.9$), $Z = -0.44$, $p = .660$. Similarly, average looking time for the last four habituation trials did not differ significantly between the upright ($Mdn = 5.2$) and inverted ($Mdn = 4.7$) conditions, $Z = -0.82$, $p = .414$.

Test Phase

Box plots of the participants' looking times during the test phase are presented in Figure 2. To determine how toddlers with WS process upright and inverted faces, looking times on the three test trials were analyzed using two Friedman tests, one for each orientation condition. Follow-up planned comparisons were conducted using Wilcoxon tests. Significantly longer looking at the switch test face compared to the familiar test face indicates holistic processing. Significantly longer looking at the novel test face compared to the familiar test face indicates that participants are able to detect the novel face as novel and that they have not become fatigued.

In the upright condition, the Friedman test indicated a significant effect for test trial type, $\chi^2(2) = 8.48$, $p = .014$. Planned comparisons indicated that as hypothesized, participants looked significantly longer during the switch test trial than during the familiar test trial, $Z = 2.29$, $p = .022$, indicating that they processed upright faces holistically. The participants also looked significantly longer during the novel test trial than during the familiar test trial, $Z = 2.97$, $p = .003$, indicating that they detected the novel test face and were not fatigued.

In the inverted condition, the Friedman test also indicated a significant effect of test trial, $\chi^2(2) = 7.11$, $p = .029$. Planned comparisons revealed that this effect was due to significantly longer looking times during the novel test trial than during the familiar test trial, $Z = 2.54$, $p = .011$, indicating that the participants were not fatigued and detected the novel test face. In contrast, as expected, looking times during the switch test trial and the familiar test trial did not differ, $Z = -0.50$, $p = .615$, indicating that the participants did not process inverted faces holistically (see Figure 2).

These findings were further supported by χ^2 analyses comparing the number of toddlers who looked longer during the switch test trial than during the familiar test trial to the number of toddlers expected to look longer during the switch test trial than the familiar test trial by chance (7; 50% of the participants). In the upright condition, 11 of 14 participants looked longer at the switch test face than at the familiar test face, $\chi^2(1) = 4.57$, $p = .033$. In contrast, in the inverted condition, only 6 of the 14 participants showed this pattern, $\chi^2(1) = 0.29$, $p = .593$. Thus, the number of participants who looked longer at the switch test face than at the familiar test face was significantly more than expected based on chance in the upright condition but not in the inverted condition.

Finally, to explore whether performance on the holistic face-processing task in either the upright or inverted orientation was related to adjusted CA or intellectual ability (Mullen ELC standard score), several analyses were conducted. First, we used Wilcoxon tests to compare the median Mullen ELC and CA of the children who looked longer at the switch test face than the familiar test face to those of the children who did not look longer at the switch test face than the familiar test face. Separate analyses were conducted for each orientation. No significant differences were found in either orientation (upright condition: exact p s > .89; inverted condition: exact p s > .53). Additionally, Spearman correlations were calculated between performance on the holistic face-processing task (switch-familiar difference scores) and both CA and ELC for each orientation. All four correlations were small (range: r ho = -.25 – .24) and none was statistically significant (p s > .39, 2-tailed). Thus, there was no evidence that either CA or ELC was related to face processing of either upright or inverted faces.

Discussion

The results of this study indicate that toddlers with WS process faces holistically when the faces are in the canonical, upright orientation. The results also provide evidence that for individuals with WS, inversion affects the holistic face processing system by toddlerhood, as toddlers with WS process inverted faces featurally. This pattern of findings supports our hypothesis that given their heightened interest in faces and their unusually extensive experience with faces from early in life, toddlers with WS would exhibit the expert-like pattern of holistic face processing that has been reported for TD infants beginning around age 7–8 months using the same paradigm (Cohen & Cashon, 2001; Ferguson et al., 2009). Our findings are the first to indicate that this perceptual expertise for faces is present in very young children with WS.

A variety of tasks has been used in past research to test holistic face processing in TD adults, children, and infants (for reviews see Maurer, Le Grand, & Mondloch, 2002; Piepers & Robbins, 2012; see also Turati, De Giorgio, Bardi, & Simion, 2010). For example, previous studies using the “composite” face paradigm, which addresses the questions of whether participants are sensitive to the holistic representation solely based on the internal features of faces, have provided evidence of holistic face processing in TD 4-year-olds (De Heering, Houthuys, & Rossion, 2007). Using a modified version of the composite face paradigm appropriate for testing infants, it has been shown that holistic face processing may be evident as early 3 months of age (Turati et al., 2010). In this study, we tested holistic processing using the switch paradigm (Cohen & Cashon, 2001), which addresses the question of whether participants are sensitive to the novel combination of familiar internal and external facial features presented in the test phase. A clear difference in participants’ ability to detect this novel combination was found based on the orientation of the face stimuli. We interpret these findings as evidence that toddlers with WS process faces holistically when the faces are upright but not when the faces are inverted. Future research should be conducted using other measures of holistic (and configural) face processing to determine how robust the effect of inversion is on the face processing system in toddlers with WS.

Although different patterns of looking time were found for upright and inverted faces during the test phase (i.e., in the familiar versus switch test trial comparisons), a statistically significant difference between the two orientations was not found in any of the following habituation phase comparisons: (1) median number of habituation trials needed to meet the habituation criterion, (2) median looking time on the first four habituation trials, or (3) median looking time on the last four habituation trials. To the extent that the duration of looking time on the first four habituation trials can be used as a measure of initial interest,

there is no evidence that our participants preferred the upright or inverted face stimuli. Based on the test trial data, however, there is clear evidence that they processed upright and inverted faces differently.

These results are consistent with previous findings with 7-month-old TD infants, which also showed an effect of inversion during the test phase but not during the habituation phase (Cohen & Cashon, 2001). Specifically, Cohen and Cashon (2001) found no difference in the mean number of trials needed to reach habituation criterion or in the average time spent looking during the first four habituation trials based on orientation. Cohen and Cashon interpreted their lack of difference by orientation finding as indicating that infants were initially equally interested in upright and inverted faces, but as the test data show, they processed them differently. Our findings and interpretation are similar.

The results of the present study are consistent with those reported for adolescents and adults with WS by Tager-Flusberg et al. (2003). However, they differ from those reported for 4 – 12-year-olds with WS by Annaz et al. (2009). The reason for the differences may be procedural. The procedures used in the present study and in the study by Tager-Flusberg et al. included a memory component. In contrast, the procedure used by Annaz et al. did not include a memory component, which allowed for online comparisons to be made. Richler et al. (2011) have argued that the simultaneous presentation of target and test faces induces an overreliance on featural face processing. This may account for the lack of an inversion effect found by Annaz et al., providing an alternate interpretation of their findings. In particular, their finding that children with WS processed both inverted and upright faces featurally could be a result of the researchers' use of a procedure that did not include a memory component.

It is important to note that although we were able to determine whether toddlers with WS are sensitive to the combination of internal and external facial features of upright and inverted faces using the habituation switch paradigm, we cannot say conclusively to which features or set of features the participants attended. To answer this question, studies incorporating eye-tracking technology will be crucial.

Two important questions that remain are 1) whether the onset of the expert pattern of holistic processing of faces is delayed for children with WS and 2) whether the earlier developmental stages of face processing found in TD infants also are followed by infants with WS or if their progression to the expert pattern takes a different path. A pattern of development including at least three steps is followed by TD infants, who evidence featural processing of both upright and inverted faces at age 3 months, then evidence holistic processing of both upright and inverted faces at age 4 months, and finally evidence the expert pattern – holistic processing of upright faces but featural processing of inverted faces – at age 7 months (Cashon & Cohen, 2004; Cohen & Cashon, 2001). Due to delays in diagnosing infants with WS who do not have serious medical problems, opportunities to test infants with WS during the first year of life are very rare. Thus far, we have only had the opportunity to test three such infants using the paradigm of the present study: an 8-month-old, a 9-month-old, and a 12-month-old. Consistent with findings for the middle step in the development of expertise for face processing by TD infants (Cashon & Cohen, 2004), all three infants with WS evidenced holistic processing of both upright and inverted faces. Many more children in this younger age range need to be tested using this paradigm, but thus far our data for the three young infants with WS, combined with the results of the present study, suggest there may be a delay in the development of the expert pattern of face processing for children with WS as measured by this task.

Our findings also are consistent with the possibility of a shared behavioral path to the development of face processing expertise for both children in the general population and children with WS. The present findings clearly show that toddlers with WS evidence holistic processing of upright faces but featural processing of inverted faces, which is the final step in the development of expert holistic processing of faces by infants in the general population (Cashon & Cohen, 2004; Cohen & Cashon, 2001; Ferguson et al., 2009). To address the possibility that infants and toddlers with WS also follow the same first two behavioral steps to the development of face processing expertise as do TD infants, a study of a larger sample of infants with WS using this paradigm is needed. Confirmation, however, would not necessarily indicate that the same neural substrate and functional processing underlie either the development of perceptual face expertise or the expert pattern of holistic face processing for children with WS as for TD children. Developmental neuroimaging studies that address these questions and identify possible alternate paths to the development of face processing expertise are critical.

Infants and toddlers with WS have developmental delay, yet they show extreme interest in people and faces from a very early age. The topic of how individuals with WS process faces has been hotly debated. Moreover, the youngest children included in prior face-processing studies were 4 years old, providing no information regarding the early development of face processing. The present findings provide the first evidence that young toddlers with WS both process upright faces holistically and show an inversion effect. These findings are an important step in understanding the developmental origins of how individuals with WS perceive and process human faces. To fully understand the nature of face processing by infants and toddlers who have WS, including the neural pathways and the effects of the specific genes deleted, many more studies are needed. Studies using the habituation “switch” paradigm focused on infants and toddlers with other genetic disorders would allow for determination of the generality of the availability of the expert pattern of holistic face processing for very young children with developmental disabilities.

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References

- Annaz D, Karmiloff-Smith A, Johnson MH, Thomas MS. A cross-syndrome study of the development of holistic face recognition in children with autism, Down syndrome, and Williams syndrome. *Journal of Experimental Child Psychology*. 2009; 102:456–486.10.1016/j.jecp.2008.11.005 [PubMed: 19193384]
- Bellugi U, Lichtenberger L, Jones W, Lai Z, St George M. The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*. 2000; 12:7–29. [PubMed: 10953231]
- Cashon CH, Cohen LB. Beyond U-shaped development in infants' processing of faces: An information-processing account. *Journal of Cognition and Development*. 2004; 5:59–80.10.1207/s15327647jcd0501_4
- Cashon CH, Ha O, Allen CL, Barna AC. A U-shaped relation between sitting ability and upright face processing in infants. *Child Development*. 2012.10.1111/cdev.12024
- Cohen, LB.; Atkinson, DJ.; Chaput, HH. *Habit X: A new program for obtaining and organizing data in infant perception and cognition studies (Version 1.0)*. Austin: University of Texas; 2004.
- Cohen LB, Cashon CH. Do 7-month-old infants process independent features or facial configurations? *Infant and Child Development*. 2001; 10:83–92.10.1006/jecp.2000.2602

- De Heering A, Houthuys S, Rossion B. Holistic face processing is mature at 4 years of age: Evidence from the composite face effect. *Journal of Experimental Child Psychology*. 2007; 96:57–70.10.1016/j.jecp.2006.07.001 [PubMed: 17007869]
- De Heering A, Rossion B, Maurer D. Developmental changes in face recognition during childhood: Evidence from upright and inverted faces. *Cognitive Development*. 2012; 27:17–27.10.1016/j.cogdev.2011.07.001
- Doyle TF, Bellugi U, Korenberg JR, Graham J. “Everybody in the world is my friend.” Hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics*. 2004; 124A:263–273.10.1002/ajmg.a.20416 [PubMed: 14708099]
- Farah MJ, Tanaka JW, Drain HM. What causes the face inversion effect? *Journal of Experimental Psychology. Human Perception and Performance*. 1995; 21:628–634. Retrieved from <http://www.ncbi.nlm.nih.gov/pubmed/7790837>. [PubMed: 7790837]
- Ferguson KT, Kulkofsky S, Cashon CH, Casasola M. The development of specialized processing of own-race faces in infancy. *Infancy*. 2009; 14:263–284.10.1080/15250000902839369
- Freire A, Lee K, Symons LA. The face-inversion effect as a deficit in the encoding of configural information: Direct evidence. *Perception*. 2000; 29:159–170.10.1068/p3012 [PubMed: 10820599]
- Gauthier I, Nelson CA. The development of face expertise. *Current Opinion in Neurobiology*. 2001; 11:219–224. [PubMed: 11301243]
- Hillier LW, Fulton RS, Fulton LA, Graves TA, Pepin KH, Wagner-McPherson C, Wilson RK. The DNA sequence of human chromosome 7. *Nature*. 2003; 424:157–164.10.1038/nature03440 [PubMed: 12853948]
- Järvinen-Pasley A, Bellugi U, Reilly J, Mills DL, Galaburda A, Reiss AL, Korenberg JR. Defining the social phenotype in Williams syndrome: a model for linking gene, the brain, and behavior. *Development and Psychopathology*. 2008; 20:1–35.10.1017/S0954579408000011 [PubMed: 18211726]
- Jones W, Bellugi U, Lai Z, Chiles M, Reilly J, Lincoln A, Adolphs R. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*. 2000; 12(Supplement):30–46. [PubMed: 10953232]
- Karmiloff-Smith A, Thomas M, Annaz D, Humphreys K, Ewing S, Brace N, Campbell R. Exploring the Williams syndrome face-processing debate: the importance of building developmental trajectories. *Journal of Child Psychology and Psychiatry*. 2004; 45:1258–1274.10.1111/j.1469-7610.2004.00322.x [PubMed: 15335346]
- Martens MA, Wilson SJ, Reutens DC. Williams syndrome: a critical review of the cognitive, behavioral, and neuroanatomical phenotype. *Journal of Child Psychology and Psychiatry*. 2008; 49:576–608.10.1111/J.1469-7610.2008.01887.X [PubMed: 18489677]
- Maurer D, Le Grand R, Mondloch CJ. The many faces of configural processing. *Trends in Cognitive Sciences*. 2002; 6:255–260.10.1016/S1364-6613(02)01903-4 [PubMed: 12039607]
- Mervis CB, John AE. Cognitive and behavioral characteristics of children with Williams syndrome: Implications for intervention approaches. *American Journal of Medical Genetics Part C-Seminars in Medical Genetics*. 2010; 154C:229–248.10.1002/Ajmg.C.30263
- Mervis CB, Morris CA, Klein-Tasman BP, Bertrand J, Kwitny S, Appelbaum LG, Rice CE. Attentional characteristics of infants and toddlers with Williams syndrome during triadic interactions. *Developmental Neuropsychology*. 2003; 23:243–268.10.1080/87565641.2003.9651894 [PubMed: 12730027]
- Mullen, EM. *Mullen Scales of Early Learning*. Circle Pines, MN: American Guidance Service; 1995.
- Piepers DW, Robbins RA. A review and clarification of the terms “holistic,” “configural,” and “relational” in the face perception literature. *Frontiers in Psychology*. 2012 Dec.3:1–11.10.3389/fpsyg.2012.00559 [PubMed: 22279440]
- Plesa-Skwerer D, Faja S, Schofield C, Verbalis A, Tager-Flusberg H. Perceiving facial and vocal expressions of emotion in individuals with Williams syndrome. *American Journal of Mental Retardation*. 2006; 111:15–26.10.1352/0895-8017(2006)111[15:PFAVEO]2.0.CO;2 [PubMed: 16332153]

- Riby DM, Hancock PJB. Viewing it differently: Social scene perception in Williams syndrome and autism. *Neuropsychologia*. 2008; 46:2855–2860.10.1016/j.neuropsychologia.2008.05.003 [PubMed: 18561959]
- Riby DM, Hancock PJB. Do faces capture the attention of individuals with Williams syndrome or autism? Evidence from tracking eye movements. *Journal of Autism and Developmental Disorders*. 2009a; 39:421–431.10.1007/s10803-008-0641-z [PubMed: 18787936]
- Riby D, Hancock PJB. Looking at movies and cartoons: eye-tracking evidence from Williams syndrome and autism. *Journal of Intellectual Disability Research*. 2009b; 53:169–181.10.1111/j.1365-2788.2008.01142.x [PubMed: 19192099]
- Riby, DM.; Porter, MA. Williams syndrome. In: Holmes, J., editor. *Advances in Child Development and Behavior: Developmental Disorders and Interventions*. Vol. 39. Burlington: Elsevier; 2010. p. 163-209.
- Richler JJ, Cheung OS, Gauthier I. Holistic processing predicts face recognition. *Psychological Science*. 2011; 22:464–471.10.1177/0956797611401753 [PubMed: 21393576]
- Rose FE, Lincoln AJ, Lai Z, Ene M, Searcy YM, Bellugi U. Orientation and affective expression effects on face recognition in Williams syndrome and autism. *Journal of Autism and Developmental Disorders*. 2007; 37:513–522.10.1007/s10803-006-0200-4 [PubMed: 16906460]
- Rossion B. Picture-plane inversion leads to qualitative changes of face perception. *Acta Psychologica*. 2008; 128:274–89.10.1016/j.actpsy.2008.02.003 [PubMed: 18396260]
- Rossion B. Distinguishing the cause and consequence of face inversion: the perceptual field hypothesis. *Acta Psychologica*. 2009; 132:300–12.10.1016/j.actpsy.2009.08.002 [PubMed: 19747674]
- Rossion B, Gauthier I. How does the brain process upright and inverted faces? *Behavioral and Cognitive Neuroscience Reviews*. 2002; 1:63–75.10.1177/1534582302001001004 [PubMed: 17715586]
- Tager-Flusberg H, Plesa-Skwerer D, Faja S, Joseph RM. People with Williams syndrome process faces holistically. *Cognition*. 2003; 89:11–24.10.1016/S0010-0277(03)00049-0 [PubMed: 12893122]
- Tanaka JW, Farah MJ. Parts and wholes in face recognition. *The Quarterly Journal of Experimental Psychology. A. Human Experimental Psychology*. 1993; 46:225–245. Retrieved from <http://www.ncbi.nlm.nih.gov/pubmed/8316637>.
- Turati C, Di Giorgio E, Bardi L, Simion F. Holistic face processing in newborns, 3-month-old infants, and adults: Evidence from the composite face effect. *Child Development*. 2010; 81:1894–1905.10.1111/j.1467-8624.2010.01520.x/full [PubMed: 21077871]
- Valentine T. Upside-down faces: A review of the effect of inversion upon face recognition. *British Journal of Psychology*. 1988; 79:471–491.10.1111/j.2044-8295.1988.tb02747.x [PubMed: 3061544]

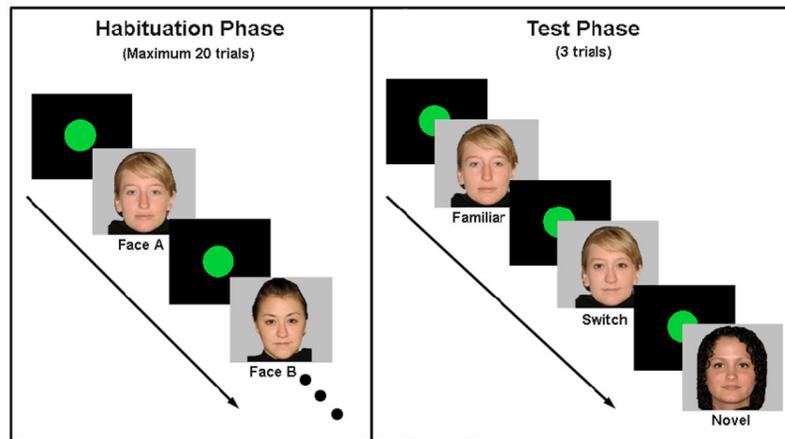


Figure 1.

Examples of stimuli presented in habituation and test phases. *Note.* Adapted from “Sitting pp is hard to do: A U-shaped relation between sitting ability and holistic face processing in infants,” by C. H. Cashon, O. Ha, C. L. Allen, and A. C. Barna (2012), *Child Development*. doi:10.1111/cdev.12024. Copyright 2012 by Wiley and Sons, Inc. Reprinted with permission.

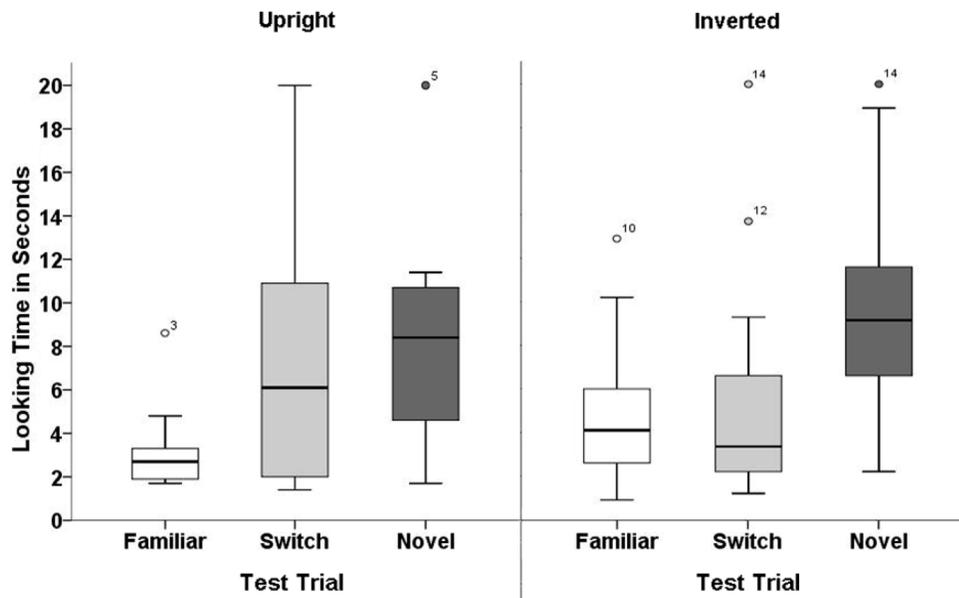


Figure 2.

Box plots representing looking times during upright and inverted test trials. The bottom of the box represents the 25th percentile (Q1), the center line the 50th percentile, and the top line the 75th percentile (Q3). Whiskers and extreme values were computed using Tukey's hinges method. Extreme values, indicated with circles, are scores that are more than 1.5 times the interquartile range (Q3-Q1) above Q3 or more than 1.5 times the interquartile range below Q1.