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Just another face in the crowd: Evidence for decreased detection of angry faces in children with Williams syndrome

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ABSTRACT

The detection of social threat is crucial for adaptive behaviour. Previous studies have shown that angry faces capture attention and are processed more efficiently than happy faces. While this *anger superiority* effect has been found in typical and atypical development, it is unknown whether it exists in individuals with Williams syndrome (WS), who show reduced social fear and atypical sociability. In this study, children with WS searched for angry or happy target faces surrounded by 2, 5 or 8 distracters (happy or angry faces, respectively). Performance was compared to that of mental age-matched controls. Results revealed no group differences for happy faces, however for angry faces, the WS, but not the control group, showed a significant performance decrease for the 8-distracters condition, indicating the absence of an *anger superiority* effect, in good agreement with evidence for abnormal structure and function in brain areas for social threat processing in WS.

1. Introduction

Detection of social threat has clear adaptive value (Darwin, 1872/1965) and social skills are critical for survival. There is a clear evolutionary advantage for humans who can efficiently recognize and detect social threat in their environment, since this ability allows them to anticipate danger, prepare defensive behaviour and hence to escape potentially dangerous situations (e.g., Öhman, Lundqvist, & Esteves, 2001; Öhman & Mineka, 2001, 2003). From early in life and throughout development, we learn to use facial expressions as powerful sources of social information, crucial for guiding behaviour towards or away from interaction.

Within this framework, a unique social-behaviour phenotype, high sociability and reduced social fear, found in individuals with Williams syndrome (WS) is of major interest. Most of these individuals are unusually friendly and show intriguing absence of fear from strangers, with whom they are generally at ease and keen to interact with (e.g., Jones et al., 2000). To date, the cognitive mechanisms underlying this consistently reported hallmark feature of WS remain unclear.

In addition to providing distinctive information about people (e.g., identity or gender), faces also transmit subtle signals related to emotion, trustworthiness, attractiveness, as well as intention. As a consequence, faces are strong facilitators for social interaction and a crucial site to convey signals of potential social threat (e.g., Blair, 2003). Faces are special stimuli for humans from the earliest stages of development. Newborns tend to look longer and to orient preferentially their attention to faces than other objects (e.g., Johnson, Dziurawiec, Ellis, & Morton, 1991). This increased and early interest for faces is thought to be related to our extraordinary abilities for processing faces and facial emotions quickly and accurately (for a review see Bruce & Young, 1986). Relative to typically developing infants, those with WS tend to overuse social engagement devices such as eye contact and to spend significantly more time focusing on faces than on objects (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Mervis & Bertrand, 1997).

Given the role of face processing skills in driving the development of social behaviour, several studies have attempted to examine face processing in relation to hypersociability in WS. While normal or near-normal levels of performance have been found in tasks for face recognition and discrimination (Bellugi, Wang, & Jernigan, 1994; Deruelle, Rondan, Livet, & Mancini, 2003; Tager-Flusberg, Plesa Skwerer, Faja, & Joseph, 2003; Udwin & Yule, 1991), some argue that these skills are both delayed (e.g., Karmiloff-Smith et al., 2004) and deviant (e.g., Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen, 1999) in WS. In addition, electrophysiological studies have shown that in spite of relatively good performance on identity tasks, brain activity to faces is abnormally organized in WS (Grice et al., 2001; Mills et al., 2000). At the neural level, however, a recent study has provided fMRI and ERP evidence of increased activation to positive emotional expressions in WS (Haas et al., 2009). In facial emotion processing, studies have shown indistinguishable performance between WS and mental age-matched groups on perception (Deruelle et al., 1999; Plesa-Skewerer, Faja, Schofield,

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Verbalis, & Tager-Flusberg, 2006) and recognition (Gagliardi et al., 2003) tasks. More recently, Santos, Milne, Rosset, and Deruelle (2007) and Santos, Rosset, and Deruelle (2009) have shown that children and adults with WS can actually categorize emotions at a higher level than that expected from their intellectual level when these are expressed in human (real and cartoon) faces.

While facial expressions of emotion, in general, provide a wealth of information, angry faces, in particular, provide specific information on potential social threat. Indeed, these are universally read as critical cues for interpersonal threat and potent warning signals. Angry faces seem thus to hold a special status for typically developing individuals and some argue that humans are biologically prepared or "hard-wired" for the recognition of anger or threat specifically (Öhman, 1993). In agreement with this, there is a great deal of evidence supporting the idea that angry faces capture attention and are processed promptly and efficiently (Vuilleumier & Schwartz, 2001). The empirical basis for specific mechanisms with regard to anger comes mostly from studies using visual search paradigms, such as the "face-in-the-crowd" task. In this task participants are instructed to detect the presence or absence of a target emotional face among a crowd of distracter faces. In a pioneering study using this task, Hansen and Hansen (1988) have shown that angry faces are detected more quickly and accurately than happy faces, independent of the number of distracters. Despite the limitations this study may have presented (e.g., Purcell, Stewart, & Skov, 1996), it has launched the idea of an *anger superiority* effect, which has received support from several further studies (e.g., Öhman et al., 2001).

Consistent with the idea that the visual system may be sensitive to the emotional valence of facial stimuli, several recent studies found more efficient search for negatively than for positively valenced face target sets among neutral distracters (e.g., Eastwood, Smilek, & Merikle, 2001; Gerritsen, Frischen, Blake, Smilek, & Eastwood, 2008), as well as more efficient search for angry target sets among happy distracters than vice-versa (e.g., Fox et al., 2000; Horstmann & Bauland, 2006). Interestingly, this *anger superiority* effect holds even for neurodevelopmental disorders, such as autism and Asperger syndrome (Ashwin, Wheelwright, & Baron-Cohen, 2006; Krysko & Rutherford, 2009), and anxiety disorders (for a review see Bar-Haim, Lamy, Pergamin, Bakermans-Kranenburg, & Van IJzendoorn, 2007).

The current study aimed at investigating whether the *anger superiority* effect can be found in children with WS, which show clinically reduced social fear and increased sociability. To this aim, we have used a "face-in-the-crowd" task and their performance was compared to that of typically developing mental age-matched controls (MA). We hypothesized that children with WS would process angry faces differently from MA controls, whereas no such group differences would be found for happy faces. Specifically, while for MA children the presence of angry, but not happy, faces should be reported with the same ease regardless of the number of distracters, for WS children the detection of both angry and happy faces should present increased difficulty with increasing number of distracters.

2. Methods

2.1. Participants

Twenty-one children with WS (12 female and 9 male) aged 10–17 years (M = 13.5; SD = 2.5) participated in this study. WS diagnosis was based on both clinical evaluation and a FISH test (fluorescent *in situ* hybridization) for microdeletion on one copy of the gene for elastin on chromosome 7. All participants fulfilled the Preus (1984) criteria for WS (e.g., characteristic facial appearance, low average birth weight, feeding difficulties, musculoskeletal problems, etc.) and were positive in the FISH test. Mental age (MA), inferred from IQ measures (WAIS-III (Wechsler, 1997) and WISC-III (Wechsler, 1996), according to the subject's age), ranged from 5 to 15 years (M = 8.3; SD = 2.6). Full-scale IQ profile (M = 60.4; SD = 12.7; range: 47–93) was characterized by a significant dissociation between verbal (M = 67.8; SD = 14.3; range: 50–101) and performance (M = 62.9; SD = 10.5; range: 46–87) IQ scores (F (1,20) = 7.62, p < .01), consistent with previous studies on WS (e.g., Jarrold, Baddeley, & Hewes, 1998). Participants were recruited via the Department of Pediatric Neurology at a local hospital (La Timone, Marseille, France) and via Regional Williams Syndrome Associations. At the time of testing, they all attended schools or specialized centres for individuals with developmental delay.

This study also included a group of 21 typically developing individuals, aged 5–15 years (M = 8.8; SD = 2.9), individually matched to WS participants on gender (12 female and 9 male) and MA (F (1,40) = .25, p > .6). Typically developing children were recruited via local schools and day-care centres and they all attended normal classes corresponding to their age level. Teachers were asked to select these children from the average level of their class thus avoiding inclusion of advanced or delayed children relative to their age. None of the participants had overt physical handicap, known neurological/psychiatric deficits or history of learning difficulties.

All participants were native French speakers and had normal or corrected-to-normal audition and vision. Informed consent was obtained for all subjects and the experimental procedure was approved by the local ethics committee.

2.2. Stimuli and procedure

Stimuli comprised cartoon faces, displaying either a happy or an angry facial expression. The same identity was used in both emotional valences. The faces consisted of coloured line drawings presented against a white background (see Fig. 1). Cartoon rather than real faces were used to avoid problems in equating real faces for confounding visual features such as shadows (e.g., Purcell et al., 1996). Importantly, Santos et al. (2009) have shown that individuals with WS have no difficulties to process emotions displayed in cartoon faces. Furthermore, previous studies using schematic faces have shown that emotional expressions are readily recognized from simple eyebrow and mouth line drawings (e.g., Magnussen, Sunde, & Dyrnes, 1994).

This study comprised two visual search tasks, examining the participants search for emotional faces in different sized arrays of faces (2, 4 or 8). Participants had either to detect the presence/absence of one angry (Task 1) or one happy face (Task 2) within a "crowd" of angry or happy faces, respectively. All trials consisted of 3 × 3 matrices (801 × 700 pixels size at the screen). In each task, half of the trials (target-present trials) consisted of one target face (discrepant face) plus other 2, 5 or 8 background faces (all similar; crowd faces). In both tasks, the target could occur at any of the 9 positions in the matrix (see Fig. 1), resulting in a total of 36 different matrices containing a target (18 target-present trials for each task, 6 trials for each matrix size). The position of the target face was also randomly assigned across trials and subjects. In the other half of the trials (36 target-absent trials, 18 trials for each task, 6 for each matrix size) all faces were similar (no discrepant face). The distance between the faces (target and the background faces) was constant across all these combinations (see Fig. 1).

Visual stimuli were presented on a 19-in. monitor connected to Macintosh laptop computer. SuperLab 4.5 experiment software (Cedrus Corporation, USA) was used to initiate trials and record responses and reaction times (RTs). Participants were tested individually in a quiet room at the Institute of Cognitive Neuroscience of the Mediterranean (CNRS, Marseille, France) or at their home. Participants were seated approximately 60 cm from the computer screen and they were asked to fixate the centre of the screen. Participants were asked to detect a discrepant face in a matrix of faces. More precisely they were to respond whether there was a happy face (Task 1) or an angry face (Task 2) in the crowd. Responses were given by pressing one of two coloured keys (red = no; green = yes) on a response box designed for use with the Macintosh laptop and the Superlab program. Before the experimental session began, all participants were presented with a practice session to familiarise themselves with the task (3 trials for each task, one corresponding to each matrix size) and to ensure they understood the instructions. Each trial consisted of a presentation of a fixation cross on an otherwise blank screen for 500 ms, followed by the matrix display and the start of trial timing. The display time was terminated by the participants' response. Half of the participants were presented with Task 1 first and the other half started with Task 2. In order to avoid interference with task instructions, the two tasks were separated by a 2-h interval, during which children watched a movie.

2.3. Data analyses

Analyses of variance (ANOVAs) with repeated measures were used for all statistical tests, and all p-values reported below were adjusted with the Greenhouse–Geisser epsilon correction for non-sphericity. The uncorrected degrees of freedom and the probability level after correction are reported. Main and/or interaction effects not significant (p > .05) are not be reported. Fisher LSD tests were used for all *post hoc* comparisons. Correlation analyses were computed using Pearson r tests. Only RTs for correct responses were considered in the analyses.

3. Results

In order to ensure that our paradigm could provide data consistent with previous studies on visual search we have first analysed