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Research report

Covert processing of facial expressions by people with Williams syndrome

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ABSTRACT

Although individuals with Williams Syndrome (WS) are empathic and sociable and perform relatively well on face recognition tasks, they perform poorly on tasks of facial expression recognition. The current study sought to investigate this seeming inconsistency. Participants were tested on a Garner-type matching paradigm in which identities and expressions were manipulated simultaneously as the relevant or irrelevant dimensions. Performance of people with WS on the expression-matching task was poor and relied primarily on facilitation afforded by congruent identities. Performance on the identity matching task came close to the level of performance of matched controls and was significantly facilitated by congruent expressions. We discuss potential accounts for the discrepant processing of expressions in the task-relevant (overt) and task-irrelevant (covert) conditions, expanding on the inherently semantic-conceptual nature of overt expression matching and its dependence on general cognitive level.

1. Introduction

Williams syndrome (WS; Williams et al., 1961) is a rare autosomal genetic disorder (estimated 1:20–25,000 live births, e.g., Kaplan et al., 2001, but see Strømme et al., 2002 for a much higher rate of occurrence), characterized by typical facial dysmorphology, renal and cardiovascular anomalies, short stature, characteristic dental malformation and hypercalcemia (McKusick, 1988). Most cases of WS are sporadic resulting from a microdeletion on chromosome 7q11.23 (but see Osborne et al., 2001). The missing region typically includes the Elastin (ELN) gene, which is hypothesized to account for the vascular and connective tissue abnormalities (Ewart et al., 1993). The other phenotypic characteristics are presumably linked to the adjacent 24 or so genes that are part of the standard deletion resulting in WS (Frangiskakis et al., 1996; Mervis et al., 1999).

IQ scores of individuals with WS are generally within the 50–70 range with a verbal IQ that is typically higher than performance IQ (Mervis et al., 1999; for a recent review and meta-analysis see Martens et al., 2008). Although children with WS are late talkers their vocabulary is relatively well developed and grammar seems to be an area of relative strength (but see Karmiloff-Smith et al., 1997 and Volterra et al., 1996 for a different view). Deficits are nevertheless seen in semantic and pragmatic aspects of language. In contrast to

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language, severe deficits in the spatial domain, particularly in visual–spatial construction tasks, are characteristic of this syndrome (Bihrle, 1990; Mervis et al., 1999).

Individuals with WS are often described as ''people-oriented''. They are hyper-sociable, exhibiting high sensitivity, empathy, and quick and spontaneous reactions to changes in the mood of other people (for a review see Doyle et al., 2004). As babies they have a tendency to stare at people's faces and to approach strangers indiscriminately (e.g., Bellugi et al., 1999; Klein-Tasman and Mervis, 2003). Yet, individuals with WS also experience high anxiety levels in social contexts along with poor performance on Theory of Mind tasks (Einfeld et al., 1997; Laws and Bishop, 2004; Tager-Flusberg and Sullivan, 2000). They have difficulties in understanding social cues and in maintaining long-term friendships.

Individuals with WS perform poorly on tasks of facial expression recognition. They are at the level of mental age controls on the Animated Full Facial Expression Comprehension Test (AFFECT; Gagliardi et al., 2003), on the Diagnostic Analysis of Nonverbal Accuracy Scale (DANVA-2; Nowicki and Duke, 1994) and on the Revised Eye Test (Baron-Cohen et al., 2001; Plesa-Skwerer et al., 2006a, 2006b). Interestingly, the deficits of individuals with WS in reading facial expressions stand in intriguing contrast to their relatively preserved face recognition ability as evident in standardized tests such as the Benton Face Recognition Test (BFRT; Benton et al., 1983, 1994), where they perform at, or close to age-appropriate level (e.g., Bellugi et al., 1994; Rose et al., 2007).

The current study sought to further investigate the perception of facial expressions in people with WS with a special focus on comparing direct and indirect processing. Of particular interest was the performance of participants with WS when processing facial expressions was indirect and did not require overt categorization. Studying covert processing would allow us to begin to tease apart the semantic-conceptual demands inherent in explicit categorization of facial expressions, from the relevant perceptual processing. To address this question a Garner-type paradigm (1976) was adopted in which facial expressions and face identity were manipulated simultaneously as the task-relevant and task-irrelevant dimensions (Levy and Bentin, 2008). In that study of healthy adults, we showed that matching expressions of same identity faces was faster and more accurate than matching expressions of different identity faces. Similarly, matching identity of faces that bore the same expressions was facilitated relative to matching identities that bore different facial expressions. Since expression was irrelevant in the identity matching task, and identity was irrelevant in the expression matching task, interaction between the task-relevant and task-irrelevant dimensions was considered evidence for indirect (perhaps covert) processing of the irrelevant dimensions. Incidentally, these results are in accord with the growing body of evidence suggesting interactions between identity and expression processing (Campbell et al., 1996; Ganel and Goshen-Gottstein, 2004; Ganel et al., 2005; Jackson et al., 2009; Schweinberger and Soukup, 1998; Schweinberger et al., 1999). Will people with WS process the task-irrelevant expressions and exhibit a similar pattern of facilitation?

2. Method

2.1. Participants

2.1.1. Individuals with WS

Sixteen individuals with a confirmed genetic diagnosis of WS [positive Fluorescent In Situ Hybridization (FISH) test], mean chronological age 17.6 (range: 12;0–26;5 years) participated in the study. Their mean verbal mental age (VMA) based on the vocabulary sub-test of the WISC-R (Hebrew version, Cahan, 1998) was 8 (range: 5–13 years; Standard Deviation – SD 2.2). Participants were recruited with the help of the Israeli WS Association. Participants or their guardians signed an informed consent as required by the Hebrew University ethical committee.

2.1.2. Controls matched on the BFRT

Sixteen children were individually matched to the participants with WS according to BFRT age-equivalence, as derived from the raw scores achieved by the participants with WS (Benton et al., 1983; see Gagliardi et al., 2003 and Karmiloff-Smith et al., 2004 for a similar procedure). The adequacy of this matching was assessed as described in Section 3.1 below. We shall refer to this group as Benton Raw Scores (BRS). Participants were typically-developing (TD), had no known cognitive, learning or developmental problems. Mean chronological age of the BRS group was 13;5, range: 6;5–27;0. The wide age range reflected the variability in performance on the BFRT seen in participants with WS.

2.1.3. VMA controls

Since expression recognition tasks may have a semantic component, VMA controls were chosen. Sixteen TD children, mean age 8 (6;0–12;3; SD: 1;9) with no known cognitive, learning or developmental problems were individually matched to the WS group. Participants and controls were matched on raw scores of the verbal sub-test of the Hebrew version of the WISC-R (Cahan, 1998). We shall refer to this group as VMA.

One-way analysis of variance (ANOVA) showed significant chronological age differences between the groups [$F(2,45) = 19.601$, $MSe = 359.1$, $p < .0001$]. Post hoc analysis revealed that both the WS and the BRS groups were older than the VMA group (WS-VMA: $p < .0001$; VMA-BRS: $p < .005$, respectively), and the WS group was somewhat older than the BRS group [WS-BRS: $p < .05$]. Along with the experimental tasks, participants and controls were tested on the short version of the BFRT and the DANVA-2 (Nowicki and Duke, 1994) to establish their level of performance on these normative tests.

2.2. Testing materials

2.2.1. BFRT (Benton et al., 1983)

The BFRT was selected as a dimension for matching face identity because, notwithstanding its questionable validity in the diagnosis of prosopagnosia (Duchaine and Nakayama, 2004), it is a well standardized test of face recognition. In this test black and white portraits of unfamiliar people with neutral expressions serve as stimuli. The short version of this