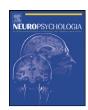
ELSEVIER

Contents lists available at ScienceDirect

### Neuropsychologia

journal homepage: www.elsevier.com/locate/neuropsychologia



## Affiliative behavior in Williams syndrome: Social perception and real-life social behavior

Anna Järvinen-Pasley<sup>a,\*</sup>, Ralph Adolphs<sup>b</sup>, Anna Yam<sup>a</sup>, Kiley J. Hill<sup>a</sup>, Mark Grichanik<sup>a</sup>, Judy Reilly<sup>c,d</sup>, Debra Mills<sup>e</sup>, Allan L. Reiss<sup>f</sup>, Julie R. Korenberg<sup>g</sup>, Ursula Bellugi<sup>a</sup>

- <sup>a</sup> The Salk Institute for Biological Studies, La Jolla, CA, USA
- <sup>b</sup> California Institute of Technology, Pasadena, CA, USA
- <sup>c</sup> San Diego State University, San Diego, CA, USA
- <sup>d</sup> University of Poitiers, France
- <sup>e</sup> Bangor University, Bangor, Wales, UK
- f Stanford University School of Medicine, Stanford, CA, USA
- g The Brain Center, University of Utah, Salt Lake City, UT, USA

#### ARTICLE INFO

#### Article history: Received 24 August 2009 Received in revised form 21 March 2010 Accepted 31 March 2010 Available online 10 April 2010

Keywords: Williams syndrome Social behavior Perception Facial expression Affect

#### ABSTRACT

A frequently noted but largely anecdotal behavioral observation in Williams syndrome (WS) is an increased tendency to approach strangers, yet the basis for this behavior remains unknown. We examined the relationship between affect identification ability and affiliative behavior in participants with WS relative to a neurotypical comparison group. We quantified social behavior from self-judgments of approachability for faces, and from parent/other evaluations of real life. Relative to typical individuals, participants with WS were perceived as more sociable by others, exhibited perceptual deficits in affect identification, and judged faces of strangers as more approachable. In WS, high self-rated willingness to approach strangers was correlated with poor affect identification ability, suggesting that these two findings may be causally related. We suggest that the real-life hypersociability in WS may arise at least in part from abnormal perceptual processing of other people's faces, rather than from an overall bias at the level of behavior. While this did not achieve statistical significance, it provides preliminary evidence to suggest that impaired social-perceptual ability may play a role in increased approachability in WS.

#### © 2010 Elsevier Ltd. All rights reserved.

#### 1. Introduction

Williams syndrome (WS) is a rare neurodevelopmental disorder resulting from a hemizygous deletion of 25–30 genes on chromosome 7q11.23 (Ewart et al., 1993; Korenberg et al., 2000). In addition to several physical abnormalities (e.g., Beuren, Schulze, Eberle, Harmjanz, & Apitz, 1964; Morris & Mervis, 2000; Williams, Barratt-Boyes, & Lowe, 1961), cognitively, the majority of individuals with WS exhibit mild to moderate intellectual impairment, with an estimated mean Full-Scale IQ (FIQ) of 50–60 (Martens, Wilson, & Reutens, 2008; Mervis et al., 2000). The FIQ masks an asymmetrical profile, in which Performance IQ (PIQ) is typically lower than Verbal IQ (VIQ) (Howlin, Davies, & Udwin, 1998; Udwin & Yule, 1990). Further, an unusual profile of cognitive dissociations has emerged

for this population: while the mature neurocognitive phenotype is associated with relative strengths in processing specifically social stimuli, such as face, significant impairments are evident in general intellectual functioning, for example, in planning and problem solving, as well as in spatial and numerical cognition (e.g., Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Martens et al., 2008). Neurological studies have further shown that near-typical performance in some tasks, such as face processing, is sustained by abnormal underlying processes (e.g., Haas et al., 2009; Mobbs et al., 2004).

A highly salient behavioral feature of WS is increased sociability (e.g., Gosch & Pankau, 1994, 1997; Udwin & Yule, 1991; von Arnim & Engel, 1964; see Bellugi et al., 2007; Järvinen-Pasley et al., 2008; Jones et al., 2000; Martens et al., 2008; Mervis & Klein-Tasman, 2000; Meyer-Lindenberg, Mervis, & Berman, 2006, for reviews). However, the social profile of WS is poorly understood and appears to be beset by intriguing paradoxes. For example, although individuals with WS are highly social and socially fearless, they nevertheless suffer from significant anxiety (Dykens, 2003; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006), exhibit substantial difficulties in social adjustment, and a tendency to suf-

<sup>\*</sup> Corresponding author at: Laboratory for Cognitive Neuroscience, The Salk Institute for Biological Studies, 10010 North Torrey Pines Road, La Jolla, CA 92037-1099, USA. Tel.: +1 858 453 4100x1224: fax: +1 858 452 7052.

 $<sup>\</sup>label{lem:email} \emph{E-mail addresses:} \ pasley@salk.edu, anna.jarvinen.pasley@gmail.com (A. Järvinen-Pasley).$ 

fer from social isolation (Udwin & Yule, 1991). Although systematic studies are relatively sparse, the increased appetitive social drive in individuals with WS has been established using a number of different paradigms, including questionnaires, behavioral observations, and experiments. For example, using two standardized temperament and personality inventories, Klein-Tasman and Mervis (2003) found that high social ratings and empathy distinguished individuals with WS from controls with other developmental disabilities. Another parent report form, The Salk Institute Sociability Questionnaire (SISQ) (Doyle, Bellugi, Korenberg, & Graham, 2004; Jones et al., 2000; Zitzer-Comfort, Doyle, Masataka, Korenberg, & Bellugi, 2007) was specifically developed by Bellugi and colleagues to elucidate the features of the social drive in WS. It gathers information regarding the individual's willingness to approach both familiar and unfamiliar people, behavior in social settings, ability to remember faces and names, eagerness to please others, empathy, and the frequency with which others approach the individual. In one study, age-related changes in social behavior in children with WS, Down syndrome (DS), and typically developing (TD) controls aged one to 13 years were investigated (Doyle et al., 2004). Consistent with earlier findings from adult participants (Jones et al., 2000), whole group analyses showed that the WS group was rated significantly higher on all aspects of sociability studied than comparison groups of individuals with various neurodevelopmental disorders and typical development. Age-related analyses showed that increased sociability was evident even among the youngest children with WS, and significantly, children with WS exceeded TD controls with respect to Global sociability and Approach strangers in every age group; similar findings were also found in relation to children with DS. These findings of significantly increased sociability in WS relative to TD have also been replicated cross-culturally (Zitzer-Comfort et al., 2007).

Observational studies have shown that beginning in infancy, individuals with WS show a strong preference for social over nonsocial stimuli (Jones et al., 2000; Mervis et al., 2003; Riby & Hancock, 2008, 2009). Experimental studies have investigated the ability of individuals with WS to make judgments regarding approach from looking at photos of unfamiliar faces (Bellugi, Adolphs, Cassady, & Chiles, 1999; Frigerio et al., 2006). In the first study of this kind, Bellugi et al. (1999) presented participants with black-andwhite photographs of unfamiliar adults, which have previously been rated in terms of approachability (both positive and negative) by typical individuals. The results showed that, while both chronological age (CA)- and mental age (MA)-matched controls performed similarly, participants with WS exhibited a positive bias by rating both positively and negatively pre-judged photographs as significantly more approachable than controls while retaining approximate rank-order. Frigerio et al. (2006) extended these findings by utilizing affective face stimuli taken from Ekman and Friesen (1976) expressing anger, disgust, fear, sadness, and happiness, in addition to neutral expressions. These stimuli had also been prerated for approachability. The results showed that participants with WS rated all but the most positively pre-judged happy photographs significantly more negatively than typical controls, suggesting that the social stimuli must be positive in valence in order for individuals with WS to perceive them as approachable.

Porter, Coltheart, and Langdon (2007) tested both social-perceptual abilities and approachability ratings with the same stimuli (the Diagnostic Analysis of Nonverbal Accuracy (DANVA; Nowicki & Duke, 1994)). Additionally, an extensive neurocognitive test battery was administered, to address three possible hypotheses with regard to approach behavior in WS: (1) that it reflects amygdala dysfunction; (2) that social stimuli have increased salience for individuals with WS; or (3) that it reflects frontal lobe dysfunction. The participants included individuals with WS and DS, as well as CA- and MA-matched TD comparison groups. The results

from the affect identification task were consistent with the literature (Gagliardi et al., 2003; Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006; Plesa-Skwerer, Verbalis, Schofield, Faja, & Tager-Flusberg, 2005), by showing that while the CA-matched controls significantly outperformed all other groups, those with WS performed at a similar level to the MA-matched controls across the four emotions (happy, sad, angry, scared). The WS group outperformed those with DS. The participants' approachability ratings were analyzed twice in order to examine the effect of affect identification ability to the perception of approachability. The analysis of the data comprising only correctly identified stimulus items revealed significant between-group differences only for happy expression. The WS, DS, and CA-matched control groups performed similarly, while the MA-matched TD controls gave significantly lower ratings. When approachability ratings to all stimuli were analyzed, unlike in the studies by Bellugi et al. (1999) and Frigerio et al. (2006), CA-matched controls in this study rated the happy stimuli as significantly more approachable than did the WS and DS groups, which performed similarly. The findings were interpreted as supporting the frontal lobe hypothesis as, despite showing similar performance to the CA-matched controls in the approachability task and exhibiting non-specific affect identification deficits, the increased approachability of individuals with WS in real life (e.g., Doyle et al., 2004; Jones et al., 2000) is likely to reflect poor response inhibition.

Most recently, Martens and colleagues conducted a study linking the approachability judgments of individuals with WS and TD controls to their amygdala volumes (Martens, Wilson, Dudgeon, & Reutens, 2009). The behavioral task was the modified Adolphs Approachability Task (Bellugi et al., 1999). The behavioral results replicated those of Bellugi et al. (1999) by showing that participants with WS rated both positive and negative stimuli as significantly more approachable than the controls. Qualitatively, reports also suggested that when judging approachability, individuals with WS relied significantly less on mouth and eye regions than the controls. Interestingly, when the approachability ratings were correlated with the participants' amygdala volumes, a positive association emerged between the right amygdala volume and approachability judgments particularly for negative stimuli, for individuals with WS only.

As discussed above, many experimental measures of sociability derived from the participants with WS themselves show some unreliability or inconsistency in the literature. Given that those measures are typically quite indirect (asking about the hypothetical approachability of a face picture), the aim of the current study will be extend the previous studies (Frigerio et al., 2006; Martens et al., 2009; Porter et al., 2007) by examining the extent to which selfratings of approachability of individuals with WS converge with their behavioral tendencies in real life, as perceived by their parents. A further rationale is that some studies have showed that individuals with WS do not perceive all of the unfamiliar faces as more approachable than controls (e.g., Frigerio et al., 2006), while ample evidence suggests significantly increased approachability in WS (e.g., Järvinen-Pasley et al., 2008; Jones et al., 2000; Martens et al., 2008). This raises the question of the extent to which the approachability ratings of individuals with WS may generalize to real-life settings. Previous evidence has also suggested that the ability to identify facial expressions may be linked to approachability ratings in individuals with WS (Porter et al., 2007). Taken together, the conflicting evidence warrants further investigation of the specific relationship between the self-rated approachability and affect identification ability, as well as the ecological validity of self-ratings of approachability, in individuals with WS.

This question is of both clinical and theoretical interest as parents of individuals with WS commonly report worrying about their children placing themselves at risk for harm by approaching unfa-

# دريافت فورى ب متن كامل مقاله

## ISIArticles مرجع مقالات تخصصی ایران

- ✔ امكان دانلود نسخه تمام متن مقالات انگليسي
  - ✓ امكان دانلود نسخه ترجمه شده مقالات
    - ✓ پذیرش سفارش ترجمه تخصصی
- ✓ امکان جستجو در آرشیو جامعی از صدها موضوع و هزاران مقاله
  - ✓ امكان دانلود رايگان ۲ صفحه اول هر مقاله
  - ✔ امکان پرداخت اینترنتی با کلیه کارت های عضو شتاب
    - ✓ دانلود فوری مقاله پس از پرداخت آنلاین
- ✓ پشتیبانی کامل خرید با بهره مندی از سیستم هوشمند رهگیری سفارشات