Neuropsychologia 48 (2010) 2110–2119

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[a,*], Ralph Adolphs[b], Anna Yam[a], Kiley J. Hill[a], Mark Grichanik[a], Judy Reilly[c,d], Debra Mills[e], Allan L. Reiss[f], Julie R. Korenberg[g], Ursula Bellugi[a]

[a] The Salk Institute for Biological Studies, La Jolla, CA, USA
[b] California Institute of Technology, Pasadena, CA, USA
[c] San Diego State University, San Diego, CA, USA
[d] University of Poitiers, France
[e] 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.

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-

* 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.
E-mail addresses: pasley@salk.edu, anna.jarvinen.pasley@gmail.com (A. Järvinen-Pasley).

0028-3932/$ – see front matter © 2010 Elsevier Ltd. All rights reserved.
doi:10.1016/j.neuropsychologia.2010.03.032

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 non-social 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-and-white 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 pre-rated 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 self-ratings 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-

Table 1
Characteristics of the two participant groups (SDs in parentheses).

	Williams syndrome ($n=20$)		TD comparison group ($n=20$)	
	Mean (SD)	Range	Mean (SD)	Range
Chronological age	29.73 (11.81)	13.13–52.29	27.90 (11.17)	12.51–48.81
VIQ (WAIS/WISC)	71 (8.36)	54–83	109 (11.48)	83–123
PIQ (WAIS/WISC)	61 (8.82)	44–77	109 (10.82)	89–132
Benton Test	19.55 (3.27)	12–26	21.80 (1.64)	18–25

miliar people (Doyle et al., 2004). This design will thus allow for the examination of the relationship between self-rated approachability of individuals with WS and their real-life social tendencies, albeit indirectly, as well as the role of facial expression processing in these behaviors. Participants with WS and a TD comparison group were administered the SISQ (Jones et al., 2000), the Adolphs Approachability Task (Bellugi et al., 1999), and a task assessing the identification of facial affect. No MA-equivalent comparison group to participants with WS was included here as numerous previous studies have robustly established that, while individuals with WS are consistently rated significantly higher than MA-matched controls on all aspects of sociability as measured by the SISQ (Doyle et al., 2004; Jones et al., 2000), individuals with WS perform at a similar level to MA-matched controls in tasks of affect identification (Gagliardi et al., 2003; Plesa-Skwerer et al., 2005, 2006; Porter et al., 2007). Finally, evidence from two previous studies indicates that MA- and CA-matched controls perform similarly in various versions of the Adolphs Approachability Task (Bellugi et al., 1999; Frigerio et al., 2006), while those with WS show significantly different pattern of performance. In the study of Porter et al. (2007), between-group differences in approachability ratings only emerged for the happy expression, and MA-matched controls (mean age = 4.9 years) rated happy stimuli as significantly less approachable than the other groups. However, this result indicates that the performance profile of individuals with WS is distinctly different from that of MA-matched TD controls, and in fact is more similar to that observed in CA-matched TD participants. Moreover, it is generally agreed that the social disposition of individuals with WS is at least in part genetically based (e.g., Deutsch, Rosse, & Schwartz, 2007; Doyle et al., 2004; Meyer-Lindenberg et al., 2005), and thus is not simply a result of general developmental delay (e.g., Doyle et al., 2004; Gosch & Pankau, 1994; Jones et al., 2000; Klein-Tasman & Mervis, 2003).

Based upon the literature reviewed above, it was hypothesized that while participants with WS will exhibit increased sociability as measured both with the SISQ and the Adolphs Approachability Task, they will show impaired facial affect identification, as compared to TD. As the findings of Frigerio et al. (2006) and Porter et al. (2007) suggest that individuals with WS can discriminate people on the basis of approachability, it was hypothesized that individuals with higher facial affect identification abilities will be more discriminative of unfamiliar people, and will thus be less inclined to approach them, as reflected by their self-ratings. As anecdotal and parental evidence nevertheless suggests that individuals with WS show increased approachability towards unfamiliar people (e.g., Jones et al., 2000), it was further predicted that self-rated approachability in WS may lack ecological validity, as reflected by parental perceptions of their children's social behavior.

2. Experiment 1: parental characterization of sociability

2.1. Method

2.1.1. Participants

Twenty individuals with WS (11 males), who had completed the three experimental measures, were identified from a pre-existing database at the Salk Institute. Genetic diagnosis of WS was established using fluorescent in situ hybridization (FISH) probes for elastin (ELN), a gene invariably associated with the WS microdeletion (Ewart et al., 1993; Korenberg et al., 2000). In addition, all participants exhibited the medical and clinical features of the WS phenotype, including cognitive, behavioral, and physical features (Bellugi et al., 2000). Twenty additional participants (7 males) were specifically recruited for the TD comparison group. These individuals were pre-screened for the level of education, and those with more than 2 years of college-level education were excluded from participation. Each participant was further screened for current and past psychiatric and/or neurological problems. Participants' cognitive functioning was assessed using the Wechsler Intelligence Scale. For participants under 16 years of age, the Wechsler Intelligence Scale Children (Third Edition; WISC-III; Wechsler, 1991) was used, and for those above 16 years of age, either the Wechsler Adult Intelligence Scale (Third Edition; WAIS-III; Wechsler, 1997), or the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999), was used. Participants were also administered the Benton Test of Facial Recognition (Benton, Hamsher, de Varney, & Spreen, 1983), a perceptual face discrimination task. In addition, all participants were native English speakers, and gave written informed consent before participation. All experimental procedures complied with the standards of the Institutional Review Board at the Salk Institute for Biological Studies.

Table 1 shows the demographic characteristics of the two groups of participants. No significant between-group differences in CA ($t(38)=0.51$, $p=0.62$) were found. As expected, the groups differed in both VIQ ($t(38)=-11.97$, $p<0.001$) and PIQ ($t(38)=-15.46$, $p<0.001$). The groups did not significantly differ in the Benton test scores ($t(38)=-2.89$, $p=0.07$).

2.1.2. Stimuli and procedures

Parents or caregivers of the participants, or if not available, other family members or friends in close contact with the participant, completed the SISQ. The criteria for the respondent were at least twice-weekly interactions with the participant during the past year, and a minimum of 3 years of knowing the participant. This is a paper and pencil measure originally requiring informants to rate the participant's social behaviors using a seven-point Likert scale with low, mid, and high endpoint labels tailored to each individual item. Additionally for some items, qualitative descriptions of social behavior are requested, resulting in both quantitative and qualitative data. Questionnaire items were designed to provide an overall index of sociability (a "Global sociability" score), which is the composite score of three subscales assessing two aspects of sociability, namely approach behavior and social-emotional behavior. Items targeting social approach behavior were divided into two categories: approach toward familiar people (the sum of three items yielding an "Approach familiars" score) and approach toward unfamiliar people (the sum of five items yielding an "Approach strangers" score). The social-emotional score is the sum of four items. For more details, including discussion on psychometric properties, see Doyle et al. (2004) and Zitzer-Comfort et al. (2007). For the WS group, all questionnaires were filled in by a parent or a caregiver. For the TD group, 40% of the questionnaires were filled in by a parent or sibling, 15% by a spouse, and the remaining 45% by a friend.

3. Results

The means and standard error means for the ratings of the two groups of participants across the SISQ subscales, as well as for the Global Sociability score, in experiment 1 are shown in Fig. 1.

The data were analyzed by a repeated-measures analysis of variance (ANOVA), with Global sociability (Approach strangers/Approach familiars/Social-emotional) entered as the within-participants factor, and group (WS/TD) as the between-participants factor. This analysis revealed a significant effect of Global sociability ($F(2, 76)=66.04$, $p<0.001$); group ($F(1, 38)=16.14$, $p<0.001$); and a Global sociability by group interaction ($F(2, 76)=6.56$, $p=0.002$). Follow-up Bonferroni corrected t test analyses showed that participants with WS were rated significantly higher compared to the TD comparison group on the Approach strangers ($t(38)=4.26$, $p<0.001$) and Social-emotional ($t(38)=3.59$, $p=0.001$) subscales of the SISQ, while the groups did not differ on their ratings on the Approach familiars subscale ($t(38)=1.68$, $p=0.10$). Pair-wise comparisons showed that both

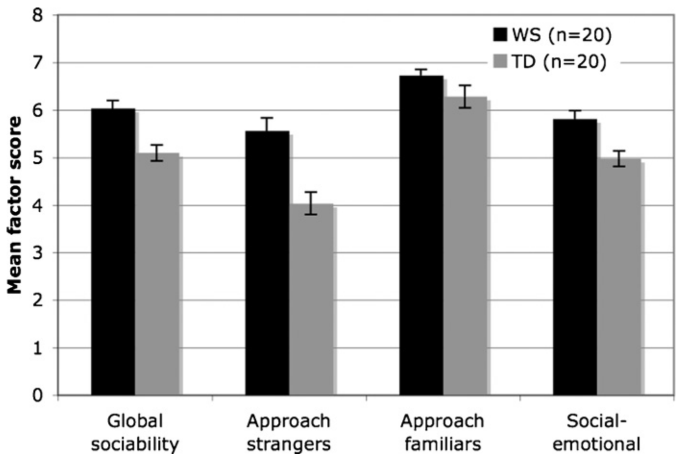

Fig. 1. Means and standard error means (±1 SEM) for the ratings of the participants with WS and their TD counterparts across the SISQ subscales, as well as for the Global Sociability score (maximum rating per scale = 7). All between-group contrasts except for Approach Familiars $p \leq 0.001$.

groups were rated higher on Approach familiars than on Approach strangers (WS: $t(19) = 5.90$, $p < 0.001$; TD: $t(19) = 8.38$, $p < 0.001$), and on Approach familiars than on Social-emotional behaviors (WS: $t(19) = 6.24$, $p < 0.001$; TD: $t(19) = 5.91$, $p < 0.001$). TD individuals were rated higher on Social-emotional behaviors than on Approach strangers ($t(19) = -4.68$, $p < 0.001$), while this was not the case for participants with WS ($t(19) = -1.12$, $p = 0.28$).

3.1. Brief discussion

The first experiment reported here sought to examine others' perceptions of sociability of individuals with WS relative to a TD comparison group. Consistent with our earlier studies of young children (Doyle et al., 2004), adolescents and adults (Jones et al., 2000), and cross-cultural samples (Zitzer-Comfort et al., 2007), the current results showed that individuals with WS were rated significantly higher on most aspects of sociability by others, as assessed by the SISQ, than TD participants. Interestingly, however, no between-group differences in ratings for the Approach familiars subscale emerged in the current study. The greatest between-group difference was evident on the Approach strangers subscale, with individuals with WS being rated significantly higher than their typical counterparts. Interestingly, the results also showed that ratings for Approach familiars were higher than ratings for Approach strangers even in the WS group (as well as in the TD group), suggesting that the social behavior of individuals with WS was not entirely indiscriminate towards others.

4. Experiment 2: self-rated approachability

4.1. Method

4.1.1. Participants

See experiment 1.

4.1.2. Stimuli and procedures

The modified version of the Adolphs Approachability Task (see Bellugi et al., 1999) included 42 black-and-white photographs of faces of unfamiliar people. Out of the original 100 stimuli (Adolphs, Tranel, & Damasio, 1998), 21 photographs that had been most consistently pre-rated positively, and 21 photographs that had been most consistently pre-rated negatively, and that together spanned as wide a range of ratings as possible, were selected as test stimuli. Participants were presented each of the stimuli individually in random order, and asked to rate them in terms of approachability using a five-point Likert scale. Participants were asked to consider how much they would like to walk up and converse with the person in the photograph. Each response was coded numerically on a scale from −2 ("No" response) to +2 ("Yes" response), and where −1 represented "Probably not" response, 0 represented "Don't know" response, and +1 represented "Maybe" response. Prior to administering the actual test stimuli, participants were familiarized with the rating scale using a training set of faces.

5. Results

Consistent with the analyses reported in Bellugi et al. (1999), Frigerio et al. (2006), and Martens et al. (2009), the participants' ratings were analyzed in relation to norms derived from the pre-ratings of the stimuli (Adolphs et al., 1998). Fig. 2 shows that, relative to the TD group, participants with WS tended to use higher approachability ratings for the positively pre-judged faces, but lower approachability ratings for most of the negatively pre-judged face stimuli. A repeated-measures ANOVA with stimulus valence (positive/negative) was entered as the within-participants factor, and group (WS/TD) as the between-participants factor, was carried out on the data. This analysis revealed significant effects of stimulus valence ($F(1, 38) = 39.71$, $p < 0.001$) and group ($F(1, 38) = 3.95$, $p = 0.05$). The stimulus valence by group interaction ($F(1, 38) = 2.54$, $p < 0.12$) was not significant, indicating that participants with WS rated all the stimuli more positively than the TD comparison group. There was a high consistency between the TD participants' ratings of approachability across the stimulus items (Cronbach's alpha = 0.89), with an intra-class correlation co-efficient of 0.86 for the negatively pre-judged faces, and 0.90 for the positively pre-judged faces. In contrast, there was low consistency in the ratings within the WS group (Cronbach's alpha = 0.38), with an intra-class correlation co-efficient of 0.24 for the negatively pre-judged faces, and −0.01 for the positively pre-judged faces. Further, in order to assess the temporal reliability of approachability judgments of typical controls with the pre-ratings of the stimuli (Adolphs et al., 1998), Cronbach's alpha was again used to calculate the intra-class correlation co-efficient for the overall responses of the typical participants. Mean Cronbach's alpha was 0.88.

5.1. Brief discussion

The second experiment reported here sought to compare self-ratings of approachability for individuals with WS relative to a TD comparison group. The results showed that participants with WS rated all stimuli more positively than the TD participants. However, the Cronbach's alpha analysis showed that there was great variability and inconsistency in the approachability ratings of individual stimulus items within the WS sample, while this was not the case for the TD comparison group. Overall, these results replicate the earlier findings of Bellugi et al. (1999) and Martens et al. (2009), which utilized the same approachability task. The current results contrast with those reported by Frigerio et al. (2006), in that participants with WS in their study only gave higher approachability ratings than the controls for the positive face stimuli, and tended to rate most of the negatively pre-judged stimuli more negatively than the TD comparison group. In the study by Porter et al. (2007), the approachability ratings of participants with WS were similar to those of the TD controls. In sum, the current results reflected a global positive bias in the self-ratings of approachability of unfamiliar faces in participants with WS. However, as in the study by Porter et al. (2007), the approachability ratings of those with WS were similar to typical when their affect identification ability was accounted for. Experiment 3 below will examine the participants' ability to decipher facially expressed affect.

6. Experiment: facial affect identification ability

6.1. Method

6.1.1. Participants

See experiment 1.

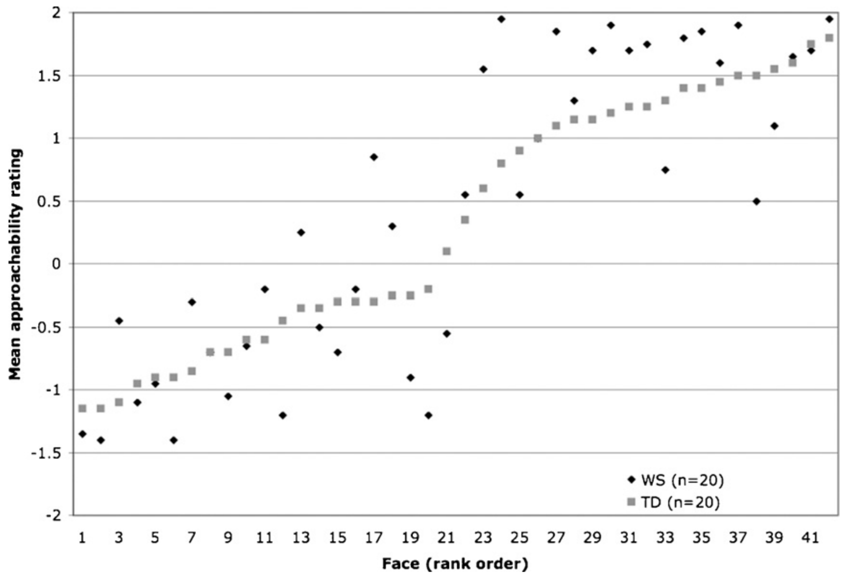

Fig. 2. Mean approachability ratings for each face stimulus by participants with WS and TD. The stimuli are rank-ordered on the *x*-axis according to their pre-judged approachability ratings. Thus, the most negative stimuli are presented at the far left, and the most positive stimuli are presented at the far right.

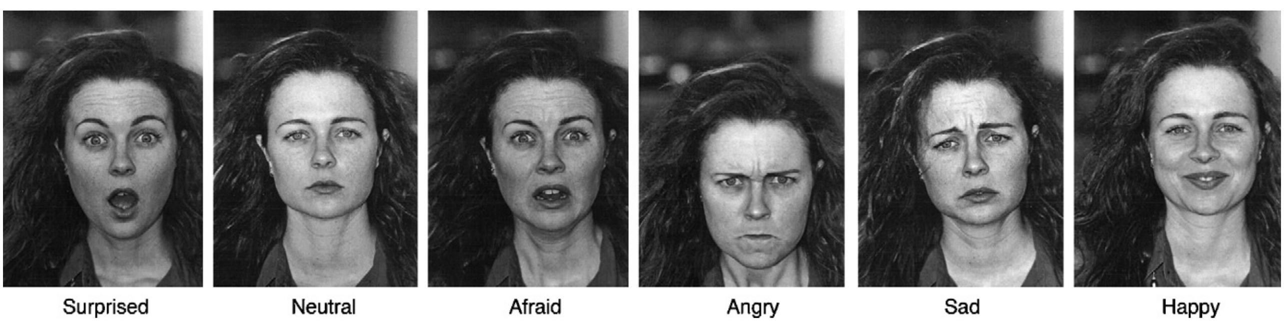

Fig. 3. Images of affective expression used in the stimuli of experiment 3.

6.1.2. Stimuli and procedures

The stimuli consist of an image board comprising six black-and-white photographs of a female face expressing extreme prototypical examples of various affective states (happy, sad, angry, afraid, surprised, and neutral; see Fig. 3). The stimuli were taken from the Affective Judgment Questionnaire for Children (Delehanty, 1993; Reilly & Delehanty, 1991). The stimuli were developed using Ekman and Friesen's Facial Action Coding System (FACS; Ekman & Friesen, 1978), and were validated on approximately 100 English-speaking TD children between the ages of 2.5 and 6.5 years (see Delehanty, 1993). The FACS is based upon the facial muscle action required to express certain emotions, and the specific muscle actions required to express certain affective states have been reliably posed, coded, and systematically measured by several investigators, most notably Ekman and Friesen. The experimenter points to the various photographs in random order and asks: "How do you think she feels in this picture?". Correctly valenced but incorrect (e.g., angry for sad) responses and vague ones (e.g., "bad" for the expression anger, sadness, and fear) are further queried with a forced-choice question that includes the target affective state and a distracter emotion. For example, "You said she feels bad here. Does she feel more angry or more sad?" Spontaneously generated correct responses were awarded two points, while correct responses to forced-choice questions were awarded one point.

7. Results

The means and standard error means for the affect-labeling scores of the two groups of participants are shown in Fig. 4.

As the data were not normally distributed, two-tailed Mann–Whitney-*U* tests were carried out on the affect identification data in order to compare performance between the two groups. This analysis showed that relative to typical individuals, participants with WS exhibited poorer ability overall ($Z = -5.45$, $p < 0.001$), with significantly lower performance with the surprised ($Z = -3.11$, $p = 0.03$), neutral ($Z = -4.98$, $p < 0.001$), and afraid ($Z = -4.45$, $p < 0.001$) stimuli.

7.1. Brief discussion

The third experiment reported here sought to assess the ability to identify facial affect in individuals with WS relative to a TD comparison group. Consistent with earlier studies (Gagliardi et al., 2003; Plesa-Skwerer et al., 2005, 2006), the current findings

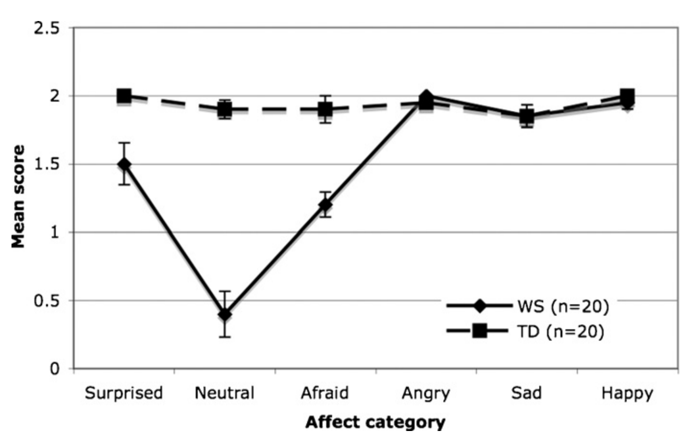

Fig. 4. Mean facial affect identification scores and standard error means (±1 SEM) for participants with WS and TD (maximum score per category = 2).

Table 2
Results from correlational analysis examining relationship between performance across experiments 1–3, for participants with WS and TD (in parentheses).

	Self-rated approachability positive faces (Exp.2)	Self-rated approachability negative faces (Exp.2)	Total self-rated approachability (Exp.2)	Affect identification (Exp.3)
Global sociability (Exp.1)	0.15 (0.57**)	−0.01 (0.59**)	0.03 (0.75**)	−0.39 (0.11)
Approach familiars (Exp.1)	−0.09 (0.19)	0.12 (0.55*)	−0.09 (0.45*)	−0.13 (0.08)
Approach strangers (Exp.1)	0.19 (0.62**)	0.10 (0.49*)	−0.15 (0.73**)	−0.34 (0.18)
Total other-rated approach (Exp. 1)	0.19 (0.61**)	0.10 (0.62**)	−0.01 (0.78**)	−0.35 (0.14)
Social-emotional (Exp.1)	−0.09 (0.24)	0.06 (0.50*)	0.02 (0.53*)	−0.05 (0.02)
Affect identification (Exp.3)	−0.63** (0.20)	0.50* (−0.07)	−0.46* (0.27)	

** $p < .01$.
* $p < .05$.

showed that individuals with WS were impaired in their overall ability to encode facial cues to affect relative to TD participants. In the current study, deficits were specifically evident with the surprised, afraid, and neutral expressions. The study by Plesa-Skwerer et al. (2005) included dynamic face stimuli with happy, sad, angry, fearful, disgusted, surprised, and neutral expressions. The findings showed that TD participants were significantly better at labeling disgusted, neutral, and fearful faces than their counterparts with WS. Similarly, the study by Gagliardi et al. (2003) included animated face stimuli exhibiting neutral, angry, disgusted, afraid, happy, and sad expressions. The results showed that participants with WS showed noticeably lower levels of performance than CA-matched controls particularly with disgusted, fearful, and sad face stimuli. The study by Plesa-Skwerer et al. (2006) only included happy, sad, angry, and afraid expressions, and found that individuals with WS showed significantly poorer performance than CA-matched controls with all but the happy expressions. In all of the above-mentioned studies, the performance of participants with WS was indistinguishable from that of MA-matched controls. Thus, the current results are largely in agreement with the previous studies showing that participants with WS show specific difficulties with identifying more complex emotions, such as surprise and fear, while the perception of the more basic emotions of happiness and anger is relatively preserved. Limitations of the current study include firstly, the free labeling paradigm, which may have resulted in participants attempting to offer actual affective labels for the neutral face stimulus; and secondly, the small number of stimuli. Although the current task was sufficiently sensitive to reveal significant between-group differences in facial affect identification ability, which are consistent with those reported in the literature, a better constructed and more comprehensive facial affect discrimination task may have revealed affect-specific patterns of correlations with the other two experimental measures; a question that future studies should address.

7.2. Relationship between performance across experiments 1–3

Table 2 shows results from the correlational analysis (Spearman's rho, two-tailed tests) comparing performance scores across experiments 1–3 for participants with WS and TD (in parentheses). Several of these correlations achieved statistical significance and we comment on them below. As an interesting relationship emerged between self-rated approachability and affect discrimination ability in the WS group, these data are presented as a scatter-plot in Fig. 5. In addition, correlations were carried out between experimental scores of interest and CA, VIQ, PIQ, and the Benton test scores. No significant correlations emerged between CA and any of the social measures (SISQ Global sociability, Approach familiars, Approach strangers, and Social-emotional; Adolphs Approachability Task self-rated approachability of positive faces, negative faces, and total approachability; Facial Affect Identification total score and individual scores for surprised, neutral, afraid, angry, sad, and happy; WS all $r < 0.32$; TD all $r < 0.40$). No significant correlation was found between Benton scores and overall facial affect identification ability for either group of participants (WS: $r(20) = 0.18$, $p = 0.45$; TD: $r(20) = 0.07$, $p = 0.78$); or between Benton scores and overall approachability ratings (WS: $r(20) = -0.32$, $p = 0.16$; TD: $r(20) = -0.12$, $p = 0.62$). For the WS group only, facial affect identification ability correlated positively with VIQ ($r(20) = 0.47$, $p = 0.04$) (TD: $r(20) = 0.15$, $p = 0.52$). PIQ did not significantly contribute to performance in either group of participants (all $r < 0.36$).

7.3. Brief discussion

Correlational analysis examining the relationship between participants' performance across experiments 1–3 revealed distinctly different patterns of association for the two groups. In the TD group, there was an excellent agreement between self- and other-ratings of approachability. More specifically, others' ratings of real-life global sociability correlated positively with self-rated approachability towards both positively and negatively valenced faces. Furthermore, self-rated approachability for both approachable- and less approachable-looking faces correlated positively with others' approachability ratings, and this correlation was particularly significant for the positively valenced faces. Others' ratings of social-emotionality correlated positively with self-ratings of willingness to approach specifically less approachable-looking faces, as well as with total self-rated approach. The social-emotional sub-

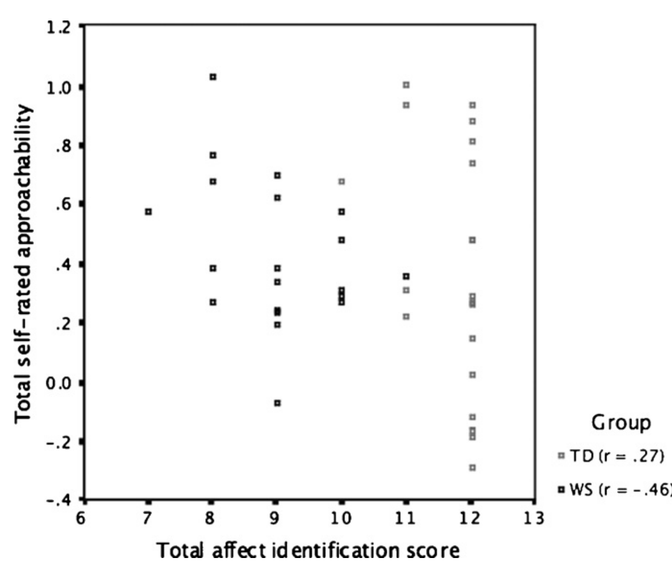

Fig. 5. Scatter-graph plotting total self-rated approachability scores (experiment 2) against total affect identification scores (experiment 3) for the WS and TD groups.

scale of the SISQ assesses aspects of social intelligence such as empathy for others and ability to remember faces and names. Thus, individuals who were perceived as high in social intelligence gave high approachability ratings for less approachable-looking faces themselves. These patterns suggest that self-rated approachability is a valid measure indexing real-life sociability in TD, and demonstrate that high sociability and approach behaviors in healthy individuals are discriminative: healthy sociable people are largely sociable only towards approachable-looking others, and those who are high in social intelligence report also approaching those who look less approachable. Likewise, the lack of correlations between self-rated approachability and affect labeling (experiments 2 and 3) bears out the conclusion that increased sociability in healthy individuals arises not from abilities related to simple facial affect identification, but at least partially from more sensitive perception and judgment of other people.

The pattern of correlations in the WS group was noticeably different from this. First, the typical correlation between self-rated approachability to faces (experiment 2) and other-rated approachability indices (experiment 1) was not significant. There are two possible interpretations from this: either individuals with WS have poor insight into their own approachability, and so their judgments of approachability in experiment 2 do not reflect how they would actually behave towards the faces they are judging; or, alternatively, they do have insight into their own approachability behavior, but their perception of the faces used in experiment 2 is impaired, such that their approachability ratings are, in essence, valid approachability judgments made about faces that they perceive incorrectly (see also Porter et al., 2007).

We favor the latter interpretation, because it is supported by additional patterns evident in Table 2. In particular, the approachability judgments of individuals with WS appear to the linked to a superficial ability to decode facial expression. By contrast, in TD, other-rated approachability appeared to be strongly linked to approachability towards approachable-looking people, and higher social-emotional intelligence. Most striking were the correlations between experiments 2 and 3. The TD group shows a lack of association between self-ratings of approachability, and facial affect identification ability, yet we found a found a highly significant negative correlation (-0.63, $p < 0.01$) between the approachability judgments for positive faces and affect identification ability in participants with WS. The poorer the individuals with WS were at judging affect in faces, the higher they rated the approachability of positive-looking faces. Similarly, the poorer they were at affect labeling, the lower they rated the approachability of negative-looking faces. Given that participants with WS were, as a group, impaired in affect labeling, these findings show that the closer to typical their affect-labeling ability, the more normal their self-rated approachability perceptions, as well as other-rated approachability, became, albeit the latter correlations failed to reach significance. This pattern of results is consistent with that reported by Porter et al. (2007). However, our data suggests that self-ratings of approachability in TD may not be linked to the construct measured by a simple affect identification task. One potential contributing factor is that these participants showed ceiling level performance in the affect-labeling task of the current study. Thus, future studies utilizing more sophisticated social-perceptual tasks are needed to validate the relationship between perceptual processing of faces and real-life sociability both in WS and in TD.

Finally, the wide age range of participants in the current study enabled us to explore the possibility that social behavior may be shaped by real-life experiences. Indeed, participants' life experience is often ignored in explanations of social cognition and social behavior, and the syndrome profile is instead seen as resulting from the genetic and neural underpinnings characterizing the syndrome. The issue of age-related changes in participants' ratings in the Adolphs Approachability Task was of specific interest, as with age, individuals with WS may accrue an increasing number of unsuccessful attempts at social relationships and history of social rejection, which, in turn, may influence their own perceptions of other people. Indeed, Gosch and Pankau (1997) found significant correlations between age and ratings of "overfriendliness" in WS in their large sample of participants, with adult participants being described as more withdrawn and less sociable than children. However, consistent with the findings of Frigerio et al. (2006) and Porter et al. (2007), showing a lack of correlations between performance in affect identification/approachability tasks and age, no association between age and self-rated approachability/others' ratings of social behavior/affect-labeling ability was found in the current study, for either group of participants. The discrepant findings described above may have at least partially arisen due to variability within the WS population with regard to social behavior, as was also demonstrated in the current study.

8. General discussion

The present experiments were designed to address the question of how self-measures of approachability are mediated by affect identification ability, and how such ratings correspond to real-life affiliative behavior, in individuals with WS. The main findings from the experiments showed that, consistent with previous data (Doyle et al., 2004; Jones et al., 2000; Zitzer-Comfort et al., 2007), relative to the TD comparison group, individuals with WS were perceived as significantly higher on aspects of sociability pertaining to approachability towards unfamiliar people and social-emotionality by their parents, as measured by the SISQ. However, an unexpected finding showed that even the participants with WS were rated as showing higher approachability towards familiar individuals than unfamiliar ones, suggesting that their approach behavior was not entirely indiscriminate. Self-ratings of approachability revealed a global positive bias in individuals with WS: they rated both the positively and negatively pre-judged photographs of faces as significantly more approachable than the TD group. This pattern of results is consistent with studies utilizing the same approachability task (Bellugi et al., 1999; Martens et al., 2009). Results from the affect identification task were also consistent with previous evidence (Gagliardi et al., 2003; Plesa-Skwerer et al., 2005, 2006; cf. Porter et al., 2007) indicating an impaired ability to recognize negative and neutral affect in individuals with WS relative to a TD comparison group.

The main finding of the current study suggested that self-ratings of approachability lack ecological validity in WS. This is the first known study to have compared self-ratings of approachability to others' perceptions of real-life approachability. At the same time, the approachability task appeared to be a valid measure of real-life approach behaviors in TD, as robust positive correlations emerged between self- and other-rated approachability. This pattern of results raises interesting questions about the apparent imbalance between a desire to approach, and actual approachability, in WS. One possibility is that it may result from the relatively artificial nature of the approachability task, i.e., photographs of faces versus real people. On this note, however, the various approach tasks used in the literature (Bellugi et al., 1999; Frigerio et al., 2006; Martens et al., 2009; Porter et al., 2007) have still successfully captured the unique feature of WS phenotype relating to increased attraction and approachability towards unfamiliar people, independent of parent ratings, which has been robustly established across a variety of measures. Thus, it appears that the performance of participants with WS in approachability tasks may be a separate issue from the real-life validity of the measure. The fact that facial affect identification ability correlated negatively with self-rated approachability in individuals with WS may be due to these tasks being similar in nature, i.e., both are perceptual and involve black-and-white photographs of faces. Consequently, this suggests that poorer affect discrimination abilities are indeed related to increased approachability perceptions in WS. At the same time, the lack of such correlations for the TD group suggested that such individuals utilized different mechanisms when making approachability judgments of faces.

A related issue concerning the processes linked to the of making approachability judgments in WS is that such individuals have been reported to rely less on the central features of faces, such as the eyes and the mouth, when making such judgments (Martens et al., 2009). Although no eye tracking was used to confirm the participants' actual attentional patterns in the Martens et al. (2009) study, this suggests a delayed developmental pattern in WS. It is thus possible that individuals with WS also utilize similar, deviant strategies when identifying facial affect, which may contribute to their impaired performance. The finding of Martens et al. (2009) appears inconsistent with results from tasks examining, for example, face matching strategies and spontaneous attention to social versus non-social stimuli, in individuals with WS (e.g., Riby, Doherty-Sneddon, & Bruce, 2008; Riby & Hancock, 2008, 2009). These studies have indicated that individuals with WS rely more on internal face features (eyes, mouth, nose) than on peripheral features (chin, hair, ears) when matching unfamiliar faces (Riby et al., 2008). In addition, eye-tracking studies have shown that participants with WS demonstrate exaggerated fixation to the eye region of the face, prolonged fixation to the face, and a significantly reduced tendency to disengage from the face, illustrating their atypically heightened interest in faces (Riby & Hancock, 2008, 2009). However, as these tasks did not require the participants to make approachability judgments, the differing nature of the tasks may explain the seemingly contradictory results. Future studies assessing eye fixation are needed to consolidate the discrepant findings across different paradigms with regard to face processing strategies in WS.

On the basis of the pattern of correlations that we found between our different dependent measures (cf. Table 2), we have suggested above that impaired perceptual processing of facial affect may be at least one contributing factor both to abnormal approachability ratings of faces, as well as potentially to increased sociability in real life, in individuals with WS. Those participants whose affect-labeling ability was the most preserved, also showed the most typical approachability ratings of faces, and were the least hypersocial in terms of approachability in their other-rated behavior. The current findings thus suggest that the increased affiliative drive characteristic of individuals with WS cannot be fully explained in terms of behavioral disinhibition (cf. Porter et al., 2007). However, an important finding showed that in TD, approachability was linked to more complex aspects of social intelligence, such as empathy, and there was no association between simple affect identification ability and approachability.

Another motivation for the current experiments was to reconcile the conflicting findings of Bellugi et al. (1999) and Martens et al. (2009), indicating a general positive perceptual bias of unfamiliar faces in WS; Frigerio et al. (2006), showing that, whereas individuals with WS perceive positively pre-judged faces as significantly more approachable, they perceive negatively pre-judged faces as significantly less approachable, than controls; and Porter et al. (2007), showing that the approachability ratings of participants with WS in response to both positive and negative stimuli did not significantly differ from those of CA-matched controls. However, this pattern of results only emerged after the stimuli, for which the participants' affect identification was impaired, was removed from the analysis. Importantly, as the task used by Porter et al. (2007) included voice and posture stimuli in addition to affective faces, their data may not be fully comparable with the previous and the current studies, which only used visual face stimuli. Our findings were comparable with those of Bellugi et al. (1999) and Martens et al. (2009). As all these studies have utilized the Adolphs Approachability Task, this suggests that it is a valid measure of self-rated approach behavior in WS. However, in the current study, there was a low consistency in the approachability ratings within the WS group, with very low intra-class correlation co-efficients for both approachable and unapproachable faces compared to the TD group. This is an indication of great variability that warrants further investigation and calls for explanations going beyond the assumption of relatively consistent syndrome profile of social cognition in WS, it being genetically determined and developmentally invariable. In fact, the findings of Porter et al. (2007) also indicated that the WS group showed the greatest variability in their approach ratings relative to all other groups, even with the stimuli which affective content they had correctly identified. Future studies should thus elucidate the sources and extent of variability in social behavior within the WS population.

The distinctiveness of the social behavior in WS has been specifically linked to their interactions with, and approachability toward, unfamiliar people (e.g., Doyle et al., 2004; Gosch & Pankau, 1997; Jones et al., 2000), to the extent that it constitutes a significant worry for parents of such individuals (Doyle et al., 2004). Several mechanisms have been proposed to contribute to this tendency. For example, as mentioned previously, researchers have noted similarities between the behavioral disinhibition characterizing both individuals with WS and patients with orbitofrontal cortical (OFC) damage (Atkinson et al., 2003; Frigerio et al., 2006). This disinhibition or drive toward social interaction is likely to be a factor independent of, and perhaps additive with, impaired affect identification. Neurobiological evidence indicates widespread anatomical abnormalities in the OFC in persons with WS, such as increased gray matter density (Eckert et al., 2006), and cytoarchitectonic abnormalities including coarse and diminished numbers of neurons, increased cell packing, and increased glia (Galaburda & Bellugi, 2000; Galaburda, Holinger, Bellugi, & Sherman, 2002). Cortical thickening is particularly apparent across areas of right frontal cortex and superior temporal sulcus in individuals with WS (Thompson et al., 2005). Instead of functional superiority, these anatomical features of WS may reflect less efficient neural packing, increased gyrification, and proportionally greater loss of white and gray matter, which may be linked to the atypical social characteristics. A recent functional magnetic resonance imaging (fMRI) study compared activation in the striatum during a response inhibition task between participants with WS and typical controls (Mobbs et al., 2007). Dysfunction in this structure has been specifically linked to behavioral disinhibition in other neurodevelopmental disorders, such as fragile X (Menon, Leroux, White, & Reiss, 2004). The findings showed that, relative to typical controls, individuals with WS exhibited significantly diminished activation in the striatum, dorsolateral prefrontal, and dorsal anterior cingulate cortices, structures that are robustly involved in behavioral inhibition in typical individuals.

In addition to the OFC, the striatum, and the cingulate cortex, other limbic structures, specifically the medial prefrontal cortex (MPFC), the insula, and the amygdala regulate emotional arousal, anxiety, endocrine function, and social impulsivity (Price, 1999). The amygdala is critical to the perception of danger, and to the subsequent regulation of appropriate behavioral responses to social-affective stimuli (Adolphs, 2003; LeDoux, 2003). Anatomically, WS is associated with a disproportionately large volume of the amygdala (Martens et al., 2009; Reiss et al., 2004). In addition, recent functional fMRI evidence indicates that WS individuals have reduced amygdala and orbitofrontal cortex (OFC) activation in response to negative face stimuli as compared to TD controls

(Meyer-Lindenberg et al., 2005). Additionally, combined event-related potentials (ERP) and fMRI evidence show that while neural responses to negative facial expressions are decreased in WS, neural activity in response to positive facial expressions is increased (Haas et al., 2009). As the fear response is regulated by the amygdala (LeDoux, 2003), and bilateral amygdala damage has been linked to deficits in fear perception and increased sense of trust toward unfamiliar people (Adolphs et al., 1998), the structural and functional abnormalities in this structure in WS might thus be associated with the heightened approachability toward strangers, and diminished perception of threat in face stimuli. In this vein, Martens et al. (2009) reported a positive correlation between right amygdala and self-rated approachability judgments particularly in response to negative faces in individuals with WS.

The current finding from experiment 3 showing impaired fear recognition in participants with WS is consistent with the hypothesis that amygdala dysfunction may contribute to their general positive bias in approachability. The findings from experiment 2 showing that individuals with WS gave increased approachability judgments to both positive and negative face stimuli relative to typical individuals may be linked to the increased neural activity in response to positive, and decreased activity to negative, face stimuli (Haas et al., 2009). Meyer-Lindenberg et al. (2005) also found that in addition to diminished amygdala activation, individuals with WS also showed a lack of OFC activation, and significantly increased activation in the MPFC, to threatening face stimuli, compared to controls. While their findings did not implicate a deficit in amygdala function per se in individuals with WS, its interactions with the prefrontal regions, specifically the OFC, appeared aberrant during the processing of threatening faces. Taken together, the neurobiological evidence reviewed above suggests that the increased social behavior characteristics of individuals with WS may be linked to the dysfunction of the OFC/amygdala (Meyer-Lindenberg et al., 2005), and the frontostriatal (Frigerio et al., 2006; Mobbs et al., 2007) systems, which may also explain the current pattern of findings.

In conclusion, the results from the present study suggest that impaired perception of affective signals from faces is one factor that contributes to impaired self-judgments of approachability from faces, as well as provide first evidence to suggest that impaired affect processing may further be linked to increased sociability in terms of approach behavior in real life. It is important to reiterate that we see impaired perception as only one component that contributes to real-life behavior, and it is likely that additional components related to motivation or disinhibitory factors play an important role as well. The current findings open a fruitful avenue for further clarifying the nature of social dysfunction characteristic of WS. More specifically, future studies should employ direct observation of social behavior, utilize sensitive social-perceptual measures, and obtain indices of past social experiences, to deepen our understanding of the factors that contribute to increased real-life social behavior in individuals with WS. More specifically, future studies should examine the relationship between participants' performance in complex social-perceptual tasks (e.g., in response to dynamic stimuli, and signals relating to approachability, such as eye gaze characteristics) and real-life approachability. We would also like to note that, as the SISQ has not previously been validated against real-life social approach behavior, the extent to which the SISQ reflects real-life social behavior remains to be further validated in future studies. Further, the use of eye tracking methodology would provide important insights into the attentional processes that are linked to the making of approachability and other socially relevant judgments in individuals WS (cf. Martens et al., 2009). This calls for the need for the development of more sensitive and reliable measures of social behavior. Insights from such studies may ultimately contribute to better-informed treatment methods in the future, which may hold the promise to enhance the social experience of individuals with WS.

Acknowledgement

This study was supported by a grant P01 HD033113-13 from the National Institute of Child Health and Human Development.

References

Adolphs, R. (2003). Cognitive neuroscience of human social behavior. *Nature Reviews Neuroscience*, 165–178.

Adolphs, R., Tranel, D., & Damasio, A. R. (1998). The human amygdala in social judgment. *Nature*, *393*, 470–474.

Atkinson, J., Braddick, O., Anker, S., Curran, W., Andrew, R., Wattam-Bell, J., et al. (2003). Neurobiological models of visuospatial cognition in children with Williams syndrome: Measures of dorsal-stream and frontal function. *Developmental Neuropsychology*, *23*, 139–172.

Bellugi, U., Adolphs, R., Cassady, C., & Chiles, M. (1999). Towards the neural basis for hypersociability in a genetic syndrome. *NeuroReport*, *10*, 1653–1657.

Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St. George, M. (2000). I. The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*, *12*(Suppl. 1), 7–29.

Bellugi, U., Järvinen-Pasley, A., Doyle, T. F., Reilly, J., Reiss, A. L., & Korenberg, J. R. (2007). Affect, social behavior, and the brain in Williams syndrome. *Current Directions in Psychological Science*, *16*, 99–104.

Benton, A. L., Hamsher, K. de. S., Varney, N. R., & Spreen, O. (1983). *Contributions to neuropsychological assessment*. New York: Oxford University Press.

Beuren, A. J., Schulze, C., Eberle, P., Harmjanz, D., & Apitz, J. (1964). The syndrome of supravalvular aortic stenosis, peripheral pulmonary stenosis, mental retardation and similar facial appearance. *American Journal of Cardiology*, *13*, 471–482.

Delehanty, S. G. (1993). *Thinking about feelings: Emotional development in the preschool years*. Unpublished Ph.D. Thesis, University of California, San Diego and San Diego State University.

Deutsch, S. I., Rosse, R. B., & Schwartz, B. L. (2007). Williams syndrome: A genetic deletion disorder presenting clues to the biology of sociability and clinical challenges of hypersociability. *CNS Spectrums*, *12*, 903–907.

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*, *124A*, 263–273.

Dykens, E. M. (2003). Anxiety, fears, and phobias in persons with Williams syndrome. *Developmental Neuropsychology*, *23*, 291–316.

Eckert, M. A., Tenforde, A., Galaburda, A. M., Bellugi, U., Korenberg, J. R., Mills, D., et al. (2006). To modulate or not to modulate? Differing results in uniquely shaped Williams syndrome brains. *NeuroImage*, *32*, 1001–1007.

Ekman, P., & Friesen, W. V. (1976). *Pictures of facial affect*. Palo Alto, CA: Consulting Psychological Press.

Ekman, P., & Friesen, W. V. (1978). *Facial action coding system (FACS): A technique for the measurement of facial action*. Palo Alto, CA: Consulting Psychologists' Press.

Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., Spallone, P., et al. (1993). Hemizygosity at the elastin locus in a developmental disorder, *Williams syndrome*. *Nature Genetics*, *5*, 11–16.

Frigerio, E., Burt, D. M., Gagliardi, C., Cioffi, G., Martelli, S., Perrett, D. I., et al. (2006). Is everybody always my friend? Perception of approachability in Williams syndrome. *Neuropsychologia*, *44*, 254–259.

Gagliardi, C., Frigerio, E., Burt, D. M., Cazzaniga, I., Perrett, D. I., & Borgatti, R. (2003). Facial expression recognition in Williams syndrome. *Neuropsychologia*, *41*, 733–738.

Galaburda, A. M., & Bellugi, U. (2000). V. Multi-level analysis of cortical neuroanatomy in Williams syndrome. *Journal of Cognitive Neuroscience*, *12*(Suppl. 1), 74–88.

Galaburda, A. M., Holinger, D. P., Bellugi, U., & Sherman, G. F. (2002). Williams syndrome: Neuronal size and neuronal-packing density in primary visual cortex. *Archives of Neurology*, *59*, 1461–1467.

Gosch, A., & Pankau, R. (1994). Social-emotional and behavioral adjustment in children with Williams-Beuren syndrome. *American Journal of Medical Genetics*, *53*, 335–339.

Gosch, A., & Pankau, R. (1997). Personality characteristics and behavioral problems in individuals of different ages with Williams syndrome. *Developmental Medicine and Child Neurology*, *39*, 327–533.

Haas, B. W., Mills, D., Yam, A., Hoeft, F., Bellugi, U., & Reiss, A. (2009). Genetic influences on sociability: Heightened amygdala reactivity and event-related responses to positive social stimuli in Williams syndrome. *Journal of Neuroscience*, *29*, 1132–1139.

Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry*, *39*, 183–189.

Järvinen-Pasley, A., Bellugi, U., Reilly, J., Mills, D. L., Galaburda, A., Reiss, A. L., et al. (2008). Defining the social phenotype in Williams syndrome: A model for linking gene, brain, and cognition. *Development and Psychopathology*, *20*, 1–35.

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., et al. (2000). II. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, *12*(Suppl. 1), 30–46.

Klein-Tasman, B. P., & Mervis, C. B. (2003). Distinctive personality characteristics of 8-, 9-, and 10-year-olds with Williams syndrome. *Developmental Neuropsychology*, *23*(1/2), 269–290.

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*(Suppl. 1), 89–107.

LeDoux, J. (2003). The emotional brain, fear, and the amygdala. *Cellular and Molecular Neurobiology*, *23*, 727–738.

Leyfer, O. T., Woodruff-Borden, J., Klein-Tasman, B. P., Fricke, J. S., & Mervis, C. B. (2006). Prevalence of psychiatric disorders in 4 to 16-year-olds with Williams syndrome. *American Journal of Medical Genetics B: Neuropsychiatric Genetics*, *141*, 615–622.

Martens, M. A., Wilson, S. J., & Reutens, D. C. (2008). Research Review: Williams syndrome: A critical review of the cognitive, behavioral, and neuroanatomical phenotype. *Journal of Child Psychology and Psychiatry*, *49*, 576–608.

Martens, M. A., Wilson, S. J., Dudgeon, P., & Reutens, D. C. (2009). Approachability and the amygdala: Insights from Williams syndrome. *Neuropsychologia*, *47*, 2446–2453.

Menon, V., Leroux, J., White, C. D., & Reiss, A. L. (2004). Frontostriatal deficits in fragile X syndrome: Relation to FMR1 gene expression. *Proceedings of the National Academy of Sciences of the United States of America*, *101*, 3615–3620.

Mervis, C. B., & Klein-Tasman, B. P. (2000). Williams syndrome: Cognition, personality, and adaptive behavior. *Mental Retardation and Developmental Disabilities Research Reviews*, *6*, 148–158.

Mervis, C. B., Robinson, B. F., Bertrand, J., Morris, C. A., Klein-Tasman, B. P., & Armstrong, S. C. (2000). The Williams syndrome cognitive profile. *Brain and Cognition*, *44*, 604–628.

Mervis, C. B., Morris, C. A., Klein-Tasman, B. P., Bertrand, J., Kwitny, S., Appelbaum, G., et al. (2003). Attentional characteristics of infants and toddlers with Williams syndrome during triadic interactions. *Developmental Neuropsychology*, *23*, 243–268.

Meyer-Lindenberg, A., Hariri, A. R., Munoz, K. E., Mervis, C. B., Mattay, V. S., Morris, C. A., et al. (2005). Neural correlates of genetically abnormal social cognition in Williams syndrome. *Nature Neuroscience*, *8*, 991–993.

Meyer-Lindenberg, A., Mervis, C. B., & Berman, K. F. (2006). Neural mechanisms in Williams syndrome: A unique window to genetic influences on cognition and behaviour. *Nature Reviews Neuroscience*, *7*, 380–393.

Mobbs, D., Garrett, A. S., Menon, V., Rose, F. E., Bellugi, U., & Reiss, A. L. (2004). Anomalous brain activation during face and gaze processing in Williams syndrome. *Neurology*, *62*, 2070–2076.

Mobbs, D., Eckert, M. A., Mills, D., Korenberg, J., Bellugi, U., Galaburda, A. M., et al. (2007). Frontostriatal dysfunction during response inhibition in Williams syndrome. *Biological Psychiatry*, *62*, 256–261.

Morris, C. A., & Mervis, C. B. (2000). Williams syndrome and related disorders. *Annual Review of Genomics and Human Genetics*, *1*, 461–484.

Nowicki, S., & Duke, M. P. (1994). Individual differences in the nonverbal communication of affect: The diagnostic analysis of nonverbal accuracy scale. *Journal of Nonverbal Behavior*, *18*, 9–35.

Plesa-Skwerer, D., Faja, S., Schofield, C., Verbalis, A., & Tager-Flusberg, H. (2006). Perceiving facial and vocal expressions of emotion in individuals with Williams syndrome. *American Journal on Mental Retardation*, *111*, 15–26.

Plesa-Skwerer, D., Verbalis, A., Schofield, C., Faja, S., & Tager-Flusberg, H. (2005). Social-perceptual abilities in adolescents and adults with Williams syndrome. *Cognitive Neuropsychology*, *22*, 1–12.

Porter, M. A., Coltheart, M., & Langdon, R. (2007). The neuropsychological basis of hypersociability in Williams and Down syndrome. *Neuropsychologia*, *45*, 2839–2849.

Price, J. L. (1999). Prefrontal cortical networks related to visceral function and mood. *Annals of the New York Academy of Sciences*, *877*, 383–396.

Reilly, J., & Delehanty, S. G. (1991). *The Affective Judgment Questionnaire for Children*. Unpublished instrument. San Diego State University, Psychology Department Psycholinguistics Laboratory.

Reiss, A. L., Eckert, M. A., Rose, F. E., Karchemskiy, A., Kesler, S., Chang, M., et al. (2004). An experiment of nature: Brain anatomy parallels cognition and behavior in Williams syndrome. *Journal of Neuroscience*, *24*, 5009–5015.

Riby, D. M., Doherty-Sneddon, G., & Bruce, V. (2008). Atypical unfamiliar face processing in Williams syndrome: What can it tell us about typical familiarity effects? *Cognitive Neuropsychiatry*, *13*, 47–58.

Riby, D. M., & Hancock, P. J. (2008). Viewing it differently: Social scene perception in Williams syndrome and autism. *Neuropsychologia*, *46*, 2855–2860.

Riby, D. M., & Hancock, P. J. (2009). Do faces capture the attention of individuals with Williams syndrome or autism? Evidence from Tracking Eye Movements. *Journal of Autism and Developmental Disorders*, *39*, 421–431.

Thompson, P. M., Lee, A. D., Dutton, R. A., Geaga, J. A., Hayashi, K. M., Eckert, M. A., et al. (2005). Abnormal cortical complexity and thickness profiles mapped in Williams syndrome. *Journal of Neuroscience*, *25*, 4146–4158.

Udwin, O., & Yule, W. (1990). Expressive language of children with Williams syndrome. *American Journal of Medical Genetics Supplement*, *6*, 108–114.

Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical Experimental Neuropsychology*, *13*, 232–244.

von Arnim, G., & Engel, P. (1964). Mental retardation related to hypercalcaemia. *Developmental Medicine and Child Neurology*, *6*, 366–377.

Wechsler, D. (1991). *Wechsler Intelligence Scale for Children—Third Edition (WISC-III)*. San Antonio, TX: Psychological Corporation.

Wechsler, D. (1997). *Wechsler Adult Intelligence Scale—Third Edition (WAIS-III)*. San Antonio, TX: Psychological Corporation.

Wechsler, D. (1999). *Wechsler Abbreviated Scale of Intelligence*. San Antonio, TX: Psychological Corporation.

Williams, J. C., Barratt-Boyes, B. G., & Lowe, J. B. (1961). Supravalvular aortic stenosis. *Circulation*, *24*, 1311–1318.

Zitzer-Comfort, C., Doyle, T. F., Masataka, N., Korenberg, J. R., & Bellugi, U. (2007). Nature and nurture: Williams syndrome across cultures. *Developmental Science*, *10*, 755–762.