Neuropsychologia 78 (2015) 159–170

Contents lists available at ScienceDirect

Neuropsychologia

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

Autonomic response to approachability characteristics, approach behavior, and social functioning in Williams syndrome

Anna Järvinen [a,*], Rowena Ng [a,b], Ursula Bellugi [a]

[a] Laboratory for Cognitive Neuroscience, The Salk Institute for Biological Studies, La Jolla, CA, USA
[b] Institute of Child Development, University of Minnesota, Twin Cities, MN, USA

ARTICLE INFO

Article history:
Received 7 February 2015
Received in revised form
6 October 2015
Accepted 8 October 2015
Available online 13 October 2015

Keywords:
Williams syndrome
Social behavior
Social functioning
Autonomic nervous system
Social interaction
Approachability task

ABSTRACT

Williams syndrome (WS) is a neurogenetic disorder that is saliently characterized by a unique social phenotype, most notably associated with a dramatically increased affinity and approachability toward unfamiliar people. Despite a recent proliferation of studies into the social profile of WS, the underpinnings of the pro-social predisposition are poorly understood. To this end, the present study was aimed at elucidating approach behavior of individuals with WS contrasted with typical development (TD) by employing a multidimensional design combining measures of autonomic arousal, social functioning, and two levels of approach evaluations. Given previous evidence suggesting that approach behaviors of individuals with WS are driven by a desire for social closeness, approachability tendencies were probed across two levels of social interaction: talking versus befriending. The main results indicated that while overall level of approachability did not differ between groups, an important qualitative between-group difference emerged across the two social interaction contexts: whereas individuals with WS demonstrated a similar willingness to approach strangers across both experimental conditions, TD individuals were significantly more willing to talk to than to befriend strangers. In WS, high approachability to positive faces across both social interaction levels was further associated with more normal social functioning. A novel finding linked autonomic responses with willingness to befriend negative faces in the WS group: elevated autonomic responsivity was associated with increased affiliation to negative face stimuli, which may represent an autonomic correlate of approach behavior in WS. Implications for underlying organization of the social brain are discussed.

1. Introduction

One powerful method for elucidating the underpinnings of human sociality is to utilize a genetically based disorder associated with altered social functioning as a model. Of particular interest to this line of investigation, Williams syndrome (WS) is a multisystem disorder (Pober, 2010), resulting from a hemizygous deletion of 25–30 genes on chromosome 7q11.23 (Ewart et al., 1993). WS is associated with a unique social phenotype saliently characterized by increased motivation for social interaction and approach (e.g., Doyle et al., 2004; Järvinen-Pasley et al., 2008; Frigerio et al., 2006), which may stem from difficulties with inhibition (Little et al., 2013). The pro-social drive of WS as reflected through a strong affinity toward unfamiliar people has been established through assorted methodologies and paradigms. At the behavioral level, such include observations (Klein-Tasman et al., 2007; Klein-Tasman and Mervis, 2003; Järvinen-Pasley et al., 2008), questionnaires (Doyle et al., 2004), eye tracking approaches (Riby and Hancock, 2008, 2009), and various experimental designs, which have, e.g., compared the willingness of individuals with WS and typically developing (TD) participants to approach strangers (e.g., Bellugi et al., 1999; Frigerio et al., 2006; Martens et al., 2009; Järvinen-Pasley et al., 2010; Martens et al., 2012). However, it is important to emphasize that despite the robustly established "hypersociability", considerable heterogeneity in multiple domains of functioning exists in WS, in e.g., cognition (perception, attention, spatial construction, social-emotional ability) (Porter and Coltheart, 2005) and social behavior (social approach tendency, response inhibition) (Little et al., 2013; Riby et al., 2014a).

Collectively, investigations employing "approachability tasks" have provided mixed findings, suggesting that approach behavior may not be entirely indiscriminate in WS. These tasks typically

* Corresponding author at: Laboratory for Cognitive Neuroscience, The Salk Institute for Biological Studies, 10010 North Torrey Pines Road, La Jolla, CA 92037-1002,USA
E-mail address: pasley@salk.edu (A. Järvinen).

http://dx.doi.org/10.1016/j.neuropsychologia.2015.10.012
0028-3932/

require participants to evaluate on a Likert-type scale how much they would like to approach a person in a facial image, which have been pre-rated for approachability characteristics (e.g., trustworthy/untrustworthy-looking; positive/negative emotional displays). Some studies have reported more positive approachability judgments in WS relative to both chronological age (CA)- and mental age (MA)-matched controls in response to both positively and negatively pre-rated faces (Bellugi et al., 1999; Jones et al., 2000; Järvinen-Pasley et al., 2010; Martens et al., 2009). By contrast, in one investigation, face stimuli displayed positive and negative standard expressions (happiness, fear, anger, disgust, sadness, neutral), and elevated approachability in WS relative to CA- and MA-matched controls was solely evident in relation to the people displaying positive emotion (Frigerio et al., 2006). In a recent study using a mouse-tracking paradigm to examine on-line trustworthiness evaluations of unfamiliar faces (Martens et al., 2012), individuals with WS relative to a CA-matched TD control group showed significantly elevated willingness to approach untrustworthy-looking people. Additionally, a set of studies have examined linkages between approachability ratings and emotion identification skills, and found that atypical approach ratings in individuals with WS were related to deficits in social perception (Järvinen-Pasley et al., 2010; Porter et al., 2007).

Extending the line of work described above, a recent study directly targeted "stranger danger" awareness and perceptions in individuals with WS using video vignettes and pre-determined questions to probe the understanding of interactions with strangers (Riby et al., 2014a). The results suggested that overall participants with WS exhibited difficulties in making trust evaluations and deciding whether to talk with the unfamiliar protagonist. The participants with WS who exhibited decreased awareness of danger also demonstrated difficulties in peer relationships and dysfunctional pro-social behavior. Another recent study reported an interesting qualitative motivational difference between individuals with WS and TD in social approach, namely, whereas high approachability in individuals with WS appeared to be driven by a desire for close interpersonal relations, TD participants demonstrated pro-social behavior with the purpose of exerting social dominance over others (Ng et al., 2014). Taken together, the evidence reviewed above suggests that while the robust appetitive social drive of individuals with WS is motivated by a desire to form relationships, inappropriate social engagement occurs at least partially because of diminished ability to socially evaluate others based on relevant characteristics and contextual cues.

Of importance here is to consider how individuals with WS may understand different types of interpersonal relationships. Using parental reports, a large-scale study by Elison et al. (2010) that included a sample of 92 adults with WS showed that approximately 30% of these individuals had no skills to form friendships and about 50% showed limited grasp of the concept of friendship. Despite this, approximately 40% of the participants were reported to enjoy good quality friendships, encompassing at least one friend of own age. While approximately 30% of the sample was described as showing adequate understanding of intimate relationships, only 12 individuals had experience of such. In line with these observations, Jawaid et al. (2011) state that, "individuals with WS experience overly problematic peer interactions and unstable relationships, despite their friendly demeanour" (p.339), and Plesa Skwerer et al. (2004) also noted that it is very rare of individuals with WS to have actual friendships, let alone a person whom to call a "best friend" (see also Gosch and Pankau (1997)). The picture of WS with respect to relationship understanding is in fact similar to that reported for individuals with other developmental disability conditions. In this vein, Jobling et al. (2000) have postulated that in case of persons with developmental disabilities, relationships are commonly misleadingly and inappropriately classified as "friendships" when they clearly fail to fulfill the concept for such. This pertains to relationships that are clearly superficial or purely instrumental involving support personnel, family friends, and facilitators. Taken together, it is clear that the majority of affected individuals do not show normal understanding of relationships, which may on the other hand be fully expected on the basis of their social-cognitive impairments encompassing the theory of mind (Tager-Flusberg and Sullivan, 2000).

Recent advances from brain-imaging studies have elucidated the neural correlates of increased approach behavior in WS, and as a result, two major hypotheses of the increased approach behavior have been proposed. First, the amygdala hypothesis postulates that alterations in the amygdala structure and/or function and its connectivity with the orbitofrontal cortex (OFC) underpin the major social features of WS (Haas et al., 2009; Meyer-Lindenberg et al., 2005; Reiss et al., 2004). The role of the amygdala in the perception of emotional facial expressions is well established (Adolphs, 2003; Herba and Phillips, 2004), and bilateral amygdala damage has been linked to atypically positive approachability judgments in response to untrustworthy-looking or negative faces (Adolphs et al., 1998). Studies of individuals with WS have reported drastically diminished amygdala activation in response to threatening faces (Meyer-Lindenberg et al., 2005) and increased activation to threatening non-social scenes (Meyer-Lindenberg et al., 2005; Thornton-Wells et al., 2011). In a similar vein, Haas et al. (2009) reported decreased amygdala activation in response to fearful faces, and increased activation to happy facial expressions, in participants with WS as contrasted with TD controls. Subsequently, it has been suggested that the amygdala dysfunction in response to threatening stimuli indexes diminished recognition of social danger, and thus is linked to the disinhibited behavior in social settings (Bellugi et al., 1999; Martens et al., 2009).

Two magnetic resonance imaging (MRI) investigations have specifically examined amygdala features in tandem with approachability tendencies in individuals with WS. First, Martens et al. (2009) related amygdala volume to approachability ratings using the Adolphs Approachability task (e.g., Bellugi et al., 1999; Jones et al., 2000; Järvinen-Pasley et al., 2010). Consistent with previous studies, individuals with WS relative to TD demonstrated increased amygdala volume and elevated approachability ratings in response to both positively and negatively pre-rated faces. Moreover, higher approachability ratings in response to negative faces were positively related right amygdala volume in the WS group, providing support to the amygdala hypothesis. In the other study, Haas and colleagues utilized the Salk Institute Sociability Questionnaire (SISQ), a parental report tapping into approach tendencies, and an implicit task testing facial expression processing in combination with functional MRI (fMRI) (Haas et al., 2010). The results showed that in individuals with WS, decreased amygdala activation to fearful facial expressions was linked to an amplified tendency to approach strangers. The authors concluded that the evidence supported the idea that abnormal amygdala response to fear is indeed associated with dysregulated social behavior in WS.

The second hypothesis posits that the increased approach behavior is underpinned by frontal lobe dysfunction resulting in impaired response inhibition. This postulation is founded upon the finding that the striatum is implicated in decision-making outcomes of social interactions related to social approval/rejection judgments. Mobbs et al. (2007) found decreased frontostriatal activation during a non-social response inhibition task and hypothesized that this may also be linked to the uninhibited social affiliation in WS, reflecting a generalized deficit. In a similar vein, Porter et al. (2007) tested participants with WS and controls on a battery comprising behavioral emotion recognition, social