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Toward a deeper characterization of the social phenotype of Williams syndrome: The association between personality and social drive

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

[a] Laboratory for Cognitive Neuroscience, the Salk Institute for Biological Studies, 10010 North Torrey Pines Road, La Jolla, CA 92037-1002, USA
[b] University of Minnesota, Twin Cities, Institute of Child Development, 51 East River Road, Minneapolis, MN 55455, USA

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ABSTRACT

Previous research has robustly established a Williams syndrome (WS) specific personality profile, predominantly characterized a gregarious, people-oriented, and tense predisposition. Extending this work, the aims of the current, cross-sectional study were two-fold: (1) to elucidate the stability of personality characteristics in individuals with WS and typically developing (TD) comparisons across development, and (2) to explore the personality attributes that may be related to the respective profiles of social functioning characterizing the two groups, which is currently poorly understood. The sample comprised of participants with WS and TD matched on chronological age. The test battery included the Multidimensional Personality Questionnaire (MPQ) and the Salk Institute Sociability Questionnaire (SISQ), an index of real-life social behavior. The main results showed that compared to the TD individuals, the WS group were consistently rated higher in Social Closeness, and this trait remained stable across development. Interpersonal behaviors were best predicted by Social Closeness in WS and by Social Potency in TD. Regression analysis highlighted that while a central motive underlying the increased drive toward social interaction in individuals with WS pertains to a desire to form affectionate relationships, TD individuals by contrast are motivated by a desire to exert social influence over others (leadership, social-dominance) and Well-Being (positive emotional disposition). In conclusion, these findings provide novel insight into social motivational factors underpinning the WS social behavior in real life, and contribute toward a deeper characterization of the WS affiliative drive. We suggest potential areas for behavioral intervention targeting improved social adjustment in individuals with WS.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder caused by a hemizygous deletion of ~28 genes on chromosome 7q11.23 (Korenberg et al., 2000). The extant literature on WS outlines a cognitive profile characterized by intriguing strengths and weaknesses, such as significantly more proficient visual–spatial processing of social (i.e., faces) over non-social stimuli (e.g., objects) (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000), higher verbal than performance IQ

* Corresponding author. Tel.: +1 8584534100x1222.
E-mail address: pasley@salk.edu (A. Järvinen).

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(Mervis & Robinson, 2000; Searcy et al., 2004), and enhanced expressive language pertaining to social-emotional qualities relative to formal linguistic competency (Reilly, Losh, Bellugi, & Wulfeck, 2004). Taken together, these features suggest enhanced processing capabilities of stimuli with specifically social relevance in individuals with WS. This pattern of functioning coexists in a context of mild to moderate intellectual impairment (Searcy et al., 2004). In addition to the distinct cognitive profile of described above, a similarly salient and distinct behavioral feature of the syndrome pertains to a unique "hypersocial" predisposition, which has been consistently documented in over two decades of the WS literature (Doyle, Bellugi, Korenberg, & Graham, 2004; Järvinen-Pasley et al., 2008; Jones et al., 2000). In this vein, WS offers an excellent context for studying human social behavior because the genetic basis is well characterized (Korenberg et al., 2000), and the hypersocial behavior is reported with striking consistency (Järvinen, Korenberg, & Bellugi, 2013).

1.1. Social profile of WS

Despite the accumulating literature, the nature of the social drive of WS remains poorly understood and systematic studies are sparse. The unusual social phenotype is characterized by an increased attraction toward faces and eyes, heightened social drive, elevated empathic concern and emotional predisposition, and diminished sensitivity to negative social information including reduced social fear (Doyle et al., 2004; Järvinen et al., 2013; Järvinen-Pasley et al., 2008; Meyer-Lindenberg et al., 2005; Riby & Hancock, 2008). The most prominent characteristic of the WS social phenotype that also appears unique to the syndrome pertains to increased approachability especially toward strangers. In order to capture dimensions of the WS social profile, Bellugi and colleagues developed a parent report form entitled The Salk Institute Sociability Questionnaire (SISQ) (Doyle et al., 2004; Jones et al., 2000; see Zitzer-Comfort, Doyle, Masataka, Korenberg, & Bellugi, 2007, for psychometric properties). Questions tap into the individual's tendencies toward approaching 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 DS (and with various other neurodevelopmental disorders, such as autism) and TD.

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). The findings showed that the individuals with WS, regardless of culture, were rated significantly higher on Approach Strangers and Global Sociability than their controls, suggesting that the WS sociability indeed is genetically determined. However, culture nevertheless had a subtle mediating effect, implicating the role of environmental factors in social behavior of WS. Empirical studies have since uncovered compelling evidence of a genetically controlled brain circuitry underpinning specific behavioral features of the social phenotype of WS (e.g., Jabbi et al., 2012; Meyer-Lindenberg et al., 2005; Meyer-Lindenberg, Mervis, & Berman, 2006; Thornton-Wells, Avery, & Blackford, 2011). Importantly, research has also suggested that affiliative tendencies in individuals with WS may be affected by their developmental stage (Järvinen-Pasley et al., 2008; Martens, Hasinski, Andridge, & Cunningham, 2012), although there is currently little systematic evidence for this.

Growing empirical evidence indicates that the WS social profile combines with distinct areas of strengths and weaknesses, revealing a new set of "paradoxes". For example, despite their "hypersocial" nature and increased drive for social interaction described above, individuals with WS show difficulties in social adjustment and social disinhibition, including problems with forming peer friendships; overfriendliness without social judgment, or pragmatic sensitivity; poor ability and/or motivation to sustain a conversation; and an inflexible, repetitive and insensitive social repertoire (Davies, Udwin, & Howlin, 1998; Klein-Tasman, Li-Barber, & Magargee, 2011; Mervis & John, 2010; Morris, 2010; Udwin, 1990). Specifically, while children with WS show competence in prosocial functions such as social motivation and social awareness, problems are apparent in social reciprocity and pragmatics (Klein-Tasman et al., 2011). It is also noteworthy that the characteristic social-emotional behavior exists in the context of other disturbed social-emotional states in WS, including a host of diagnostically significant anxiety disorders (Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006; Mervis & John, 2010; Muñoz et al., 2010) and attentional difficulties (Leyfer et al., 2006; Muñoz et al., 2010). Psychiatric research has further introduced enigmas within the cognitive and behavioral WS profile.

Our extensive studies with this population suggest that stronger cognitive skills are associated with increased anxiety in WS (Ng & Bellugi, 2013); however, individuals with WS have generally been characterized as demonstrating more intense phobic and physiological anxieties relative to TD controls and IQ-matched developmentally delayed peers, despite the fact that the clinical groups did not differ in the experiencing of general fear (Gosch & Pankau, 1997; Jawaid et al., 2011; Leyfer et al., 2006; Ng & Bellugi, 2013). Combined with the level of intellectual function, it has been suggested that the characteristic social profile of WS predisposes such individuals to social vulnerability, such as risk of social isolation, difficulties in employment, bullying, abuse, and erratic relationships (Jawaid et al., 2011). Thus, elucidating the stability and the role of personality factors in the increased social drive associated with WS may help identify areas that may be sensitive to intervention with respect to improving social adaptation in WS.