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

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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.

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Keywords

Personality; Social Behavior; Social Motivation; Social Functioning; Williams Syndrome

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 (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 et al., 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; Jawaaid 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.

1.2. Personality in WS

Research to date examining personality attributes in WS has attempted to capture the enigmatic and contradistinctive socially relevant traits. In an early study, Gosch and Pankau (1997) used the Child Behavior Checklist (CBCL) (Achenbach, 1983) to explore both temperament traits and behavioral problems associated with WS. The results revealed that while individuals with WS possessed a gregarious disposition, they experienced significant difficulties in maintaining close relationships. This finding is surprising and appears “paradoxical” in the context of evidence suggesting that the social behavior of individuals with WS, relative to other developmentally delayed (DD) populations, is particularly well characterized by the Social Closeness trait (i.e., high sociability and desire for social relations) of the Multidimensional Personality Questionnaire (MPQ) (Klein-Tasman & Mervis, 2003; Tellegen, 1985). Thus, this suggests that while individuals with WS have a strong urge to socially interact with others, they struggle with developing and maintaining friendships, as described above, which may implicate that their social relations are superficial in nature.

Specifically, Klein-Tasman & Mervis (2003) used two standardized temperament and personality questionnaires, the Children's Behavior Questionnaire (CBQ) (Rothbart, Ahadi, Hershey, & Fisher, 2003) and a parent-version of the MPQ (Tellegen, 1985) to construct an empirically derived personality profile of WS. The comparison group consisted of children with other developmental disabilities. The findings showed that the personality characteristics that clearly distinguished individuals with WS from those with other learning disability conditions with 96% sensitivity and 85% specificity included a lack of shyness, high empathy, and gregariousness. In addition, individuals with WS were uniquely people-oriented, visible, tense, and sensitive/anxious. As an intriguing extension to this study, Jabbi et al. (2012) recently reported global reduction in dorsal anterior insula volume, together with compromised connectivity between the insula, the amygdala, and the orbitofrontal cortex (OFC) in individuals with WS relative to typical controls. Further, the structural and functional alterations in the anterior insula predicted the extent to which the participants displayed the distinct WS personality characterized by hypersociability, anxiety, and empathy, i.e., composite ratings of the MPQ items people-oriented, gregarious, visible, sensitive, and tense. This was interpreted as evidence of the intricate interplay between neural systems, genetics, and complex behavioral features as responsible for human social behavior.

1.3. Social Behavior and Personality

However, personality traits that are not directly linked to social behavior have been less thoroughly explored in individuals with WS. It is possible that they may also contribute to the difficulties with interpersonal relationships experienced by such individuals (Jawaid et al., 2011). For example, difficulties with inhibitory control and negative emotionality, features that are thought to be linked to cognitive impairment, are commonly observed in

both WS and other DD populations (Tonge, 1999). Notably, the development of these personality dimensions appears atypical in WS. For example, those with WS face problems with control, attention, impulsivity and high energy, all of which have been reported to intensify with age (Gosch & Pankau, 1997; Udwin & Yule, 1991). Additionally, individuals with WS have been reported to experience greater anxiety, stress, nervousness, and fear relative to other DD groups (Dykens, 2003; Sarimski, 1997). Leyfer and co-authors (2006) reported prevalence rates of, e.g., 54% for specific phobia, 12% for generalized anxiety disorder, and 7% for separation anxiety disorder, together with only 2% for social phobia and 1% for panic disorder. In brief, personality dimensions other than socially relevant attributes may also have unique contributions to the distinct social behavior of WS; however, these two types of traits have rarely been addressed in parallel. We sought to address this gap in literature by delineating first the personality attributes that most closely associated with interpersonal behaviors in WS and TD, and subsequently, from these features, identify the attributes that best predicted social behaviors in real life.

Presently, as mentioned above, little is known about the development of the distinct WS personality. It is specifically unknown whether certain traits may appear more stable or malleable within this population, and whether others may be consistently exaggerated relative to normative personality development. Further, the relationship between the distinct personality traits in WS and the increased social drive is unclear; the current study was aimed at addressing this gap with the view of deepening the characterization of the nature of the social drive in WS. By elucidating the genetically determined expression of hypersociability in the context of personality will allow for the delineation of features of social functioning that are unique to WS. This research may help identify areas that may be sensitive to intervention with respect to improving social adaptation in WS.

1.4. The Present Study

The present exploratory study sought to systematically characterize personality features in parallel to social drive in individuals with WS and a TD comparison group matched for chronological age. The aims of this study were: (1) to examine the age-related differences in personality traits in individuals with WS and the TD comparison group by using the MPQ (Tellegen, 1985); (2) To relate the personality profiles to profiles of real life social behavior derived from the SISQ. The SISQ has been previously used as an index of social functioning in real life (Järvinen-Pasley et al., 2010); thus, the parallel study of personality features and real life social behaviors in WS extends the existing literature in new ways. Previous research has identified Social Closeness as a trait that best captures the social aspects of personality that are unique to WS in children aged 8-10 years (Klein-Tasman & Mervis, 2003); thus, a secondary aim was to compare the predictive power of this trait over other personality dimensions for social behavior and social functioning across development. We hypothesized that the traits captured by the Social Closeness construct may be closely related to social affiliative behaviors in both WS and TD. We set to explore these questions cross-sectionally at two developmental stages: child/adolescent versus adult.

2. Method

2.1. Participants

The study enrolled 63 individuals with WS (28 females; $M = 20.70$ years, $SD = 12.83$, $range = 4.73 - 51.49$ years) and 91 TD individuals (47 females; $M = 21.44$ years, $SD = 12.08$, $range = 5.62 - 51.71$ years). Data of 10 WS and 4 TD participants were excluded from the analyses due to incomplete IQ test data, failure to pass the screening criteria, or noncompliance with the study procedures. The WS and TD participants were further divided into two subgroups based on their age: *child/adolescent* (<18 years) and *adult* (>18 years), to enable the examination of age-related differences in personality and sociability (see Table 1 for participant characteristics). A total of 28 participants with WS were classified as belonging to the child/adolescent group, and the remaining 27 individuals with WS were designated to the adult group. For the TD comparison group, the child/adolescent subgroup included 34 individuals while the remaining 53 participants were designated to the adult group. Independent t-tests revealed no significant between-group differences in mean age in either the child/adolescent group, or in the adult group (both t 's < 1.81, *ns*)

The diagnosis of WS was established using the Diagnostic Score Sheet (American Academy of Pediatrics, Committee on Genetics, 2001), and the fluorescent in situ hybridization test (FISH). The TD participants were screened for exclusionary criterion, i.e., history of neurological trauma, psychiatric conditions, or developmental delays. All participants, or their caregivers or guardians, and family members provided a written informed consent and/or assent for the enrolled children. Participants and their caregivers were explained that their involvement in the study is voluntary and they were reimbursed at an hourly pecuniary rate.

Participants' cognitive functioning was assessed using the Wechsler Intelligence Scale. For participants under 16 years of age, either the Wechsler Intelligence Scale Children (WISC) - Third Edition (Wechsler, 1991) or the WISC Revised (Wechsler, 1974) was used. For those above 16 years of age, either the Wechsler Adult Intelligence Scale (WAIS) Third Edition (Wechsler, 1997), WAIS Revised (Wechsler, 1981), or the Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999) was used. However, eight of the child participants with WS were given the Kaufman Brief Intelligence Test (K-BIT; Kaufman & Kaufman, 1990) given their age (i.e., WISC are administered between age 6 to 16 year olds) or previous exposure to the WISC test. Importantly, analyses were completed with and without these participants' scores to ensure significant results are not achieved due to their data inclusion. The difference in IQ tests administered across participants is due to the fact that the participants were tested as a part of a long-standing multi-site research program, and the test battery has undergone some revisions over time. However, the Wechsler instruments (WISC-R, WAIS) have been shown to demonstrate high intra-test reliability with each other even when applied to developmentally delayed populations (Spitz, 1983), and strong test re-test reliability has been documented (Naglieri & Pfeiffer, 1983). Other Wechsler instruments were found to have factor structures that are reliable when cross-validated (i.e., WAIS-R, WISC-III)(Allen & Thorndike, 1995), and high validity (i.e., WAIS-III)(Ryan & Ward, 1999). Further, K-BIT has been documented to have high validity when measured against Wechsler

instruments for adults (Walters & Weavers, 2003), and children (Canivez, Neitzel, & Martin, 2005), and substantial correlations with WISC (Kaufman & Kaufman, 1990; Prewett, 1992) even in clinical populations (Javorsky, 1993). As expected, individuals with WS scored at a level indicative of a mild cognitive impairment in terms of VIQ, PIQ and Full Scale IQ (FIQ) measures (see Table 1). The scores of the participants with WS were significantly lower than those of TD comparison individuals on all three measures ($ts > 17.39$, $ps < .001$).

2.2. Materials

Consistent with the existing literature, the participants' personality was assessed by the MPQ-Parent Version (Tellegen, 1985; see Klein-Tasman & Mervis, 2003 for more information about the Parent Version). The MPQ was specifically selected as a reliable index of personality as it has been previously shown to successfully and sensitively discriminate individuals with WS from other DD populations (Klein-Tasman & Mervis, 2003). The inventory consists of 34 items that comprise the following 11 subscales: Absorption, Achievement, Aggression, Alienation, Control, Harm Avoidance, Social Closeness, Social Potency, Stress Reaction, Traditionalism, and Well-being. Parents were asked to rate their child on a 4-point scale, resulting in a high or a low score for each of the items: 1 - *definitely low on the trait*; 2 - *probably low on the trait*; 3 - *probably high on the trait*; 4 - *definitely high on the trait*. The Absorption subscale assesses the responsivity of the individual and the extent to which one is immersed in fantasies or stimuli. Achievement assesses the degree to which an individual is hard working, persevering, and ambitious, while the Aggression subscale assesses the participant's capacity for forgiveness and pugnacity. Alienation measures the extent to which one feels victimized or exploited, and Control assesses the degree to which one is reflective, neat and organized, and sensible. Harm Avoidance specifically taps onto safety consciousness, together with enjoyment of adventurous and thrilling activities. Social Potency indexes pleasure in leadership roles or attention seeking, visibility, dominance, and persuasive skills, while the Social Closeness subscale assesses gregariousness, and the extent to which one is people-oriented and affectionate toward others. Stress Reaction evaluates the extent to which one is even-tempered, tense, or sensitive, whereas the Well-being subscale assesses the intensity of positive emotionality and thoughts. Lastly, the Traditionalism subscale indexes the extent to which the participant adheres to conventional practices and follows orders from authorities.

The SISQ was administered to assess overt affiliative tendencies and behaviors in social settings, as well as social-emotional characteristics, of the participants. This is a paper and pencil measure 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. Questionnaire items were designed to assess two aspects of sociability, namely approach behavior and social-emotional behavior. Items targeting social approach behavior index approach toward both familiar and unfamiliar people. The social-emotional scale indexes emotional responsivity towards others, empathic tendencies, and the desire to please others (for more details, including discussion on psychometric properties, see Doyle et al., 2004; Zitzer-Comfort et al., 2007).

2.3. Procedures

To maintain consistency with the existing literature (Järvinen-Pasley et al., 2010), for the WS group, all questionnaires were filled in by a parent or a caregiver. Two participants with WS indicated that their forms were self-reported; their data were subsequently excluded from the final statistical analysis. Caregivers of the child/adolescent TD group were also requested to fill in the inventories on their child's behalf. However, as several of TD adults were living apart from family members, these participants were instead instructed to complete the inventories with the assistance of a family member or a spouse. No significant differences in SISQ and MPQ ratings were found between those TD adults who self-reported versus those who completed with a relative or spouse.

2.4. Data Analyses

Independent t-test was implemented to assess between group differences in personality factors across child/adolescents with WS versus TD comparison individuals of corresponding age; likewise, this analysis was conducted for adults with WS versus TD. To evaluate within- group developmental differences, paired t-tests were computed for the child/adolescent participants versus the adults with WS and subsequently repeated for the TD group. Notably, the central aims of the study were to clarify how personality factors differ in individuals with WS contrasted with TD, and how these factors are further associated with their respective social functioning profiles. As such, group and developmental differences in SISQ indices for those with WS and TD are not outlined, as these patterns have been assessed and consistently documented in numerous prior studies (see Doyle et al., 2004; Jarvinen-Pasley et al., 2008; Jones et al., 2000; Zitzer-Comfort et al., 2007). Pearson bivariate correlations were used to measure the associations among MPQ dimensions and SISQ indices for the WS and TD groups, informing which predictors to include in the regression analysis. Finally, hierarchical linear regressions were independently conducted to measure the predictive values of age, gender, and personality traits on SISQ Social Approach and Social Emotionality in WS and TD.

3. Results

3.1. Preliminary Analyses: Gender and IQ with Personality and Social Indices

Preliminary data analyses were carried out to assess personality differences related to gender within the WS and TD groups. Independent t-tests showed that TD female participants scored higher on Social Closeness ($M=3.33$, $SD=.70$; $t(85) = 3.08$, $p < .01$) and marginally higher on Tradition ($M=3.13$, $SD=.63$) as compared to their male counterparts ($M=2.85$, $SD=.74$; $M=3.14$, $SD=.65$; $t(85) = 1.96$, $p = .05$). No significant differences between male and female participants were observed in the WS group.

Pearson bivariate correlations indicated that IQ and personality were weakly correlated. In those with WS, VIQ was positively correlated with Alienation ($r=.35$) and Stress Reaction ($r=.28$, $ps < .05$) but not with any SISQ indices. In TD, PIQ was negatively associated with Harm Avoidance ($r=-.24$), and VIQ was positively correlated with Stress Reaction ($r=.23$) and Social-Emotionality ($r=.27$, $ps < .05$).

3.2. Personality Differences Between Participants with WS versus TD Across Development

Independent t-tests were carried out to explore differences between participants with WS versus TD across developmental periods (child/adolescents with WS versus TD; adults with WS versus TD). Table 2 displays the mean personality ratings for the WS and TD participants across the two developmental groups. Results showed that the TD participants scored significantly higher than those with WS in Achievement across both child/adolescent ($t(60) = -5.12, p < .001$) and adult ($t(78) = -7.02, p < .001$) subgroups. Although adults with WS and TD did not differ on Control, child/adolescent participants with WS scored lower in this construct relative to the TD participants of corresponding age ($t(60) = -3.69, p < .01$). Both the child/adolescent ($ts > 2.95, ps < .01$) and adult ($ts > 2.97, ps < .01$) participants with WS were rated significantly higher on Stress and Social Closeness as compared to their TD counterparts of corresponding ages. There were no other significant group differences in either higher-order or lower-order personality factors between the WS and TD participants across the two developmental stages.

3.3. Age-Related Personality Differences Within WS and TD Groups

Further t-tests were applied to explore within-group effects in personality factors across development (child/adolescent versus adult). Both WS and TD adults obtained higher scores in Achievement ($ts < -2.68, ps < .01$) as compared to the child/adolescent participants with the same group identifier. TD children/adolescents were rated higher in Stress Reaction than their adult counterparts ($t(85) = 2.14, p < .05$), although this age-related difference was not observed in participants with WS. Further, the adults with WS relative to their child/adolescent counterparts were rated higher on Control ($t(53) = -3.11, p < .01$) and marginally greater on Tradition ($t(53) = -1., p = .06$). However, no developmental differences emerged for either the Social Potency or the Social Closeness scores within either the WS or TD groups, suggesting that personality traits pertaining to sociability in both WS and TD appear consistent across development.

3.4. Correlations between Personality Profiles and Social Functioning

Two participants with WS (one child, one adult) and six TD adult comparison individuals did not fully complete the SISQ; thus, their data were excluded from the regression analyses requiring scores from both of the inventories. Correlations between the MPQ personality dimensions and the SISQ Social Approach and Social Emotionality subscales were computed (see Table 3). Social Closeness was correlated with the two SISQ constructs for both the WS and TD groups ($rs > .37, ps < .001$). Additionally, for both groups, SISQ Social Emotionality positively associated with MPQ Tradition and MPQ Well-being ($rs > .30, ps < .01$). Higher MPQ Achievement ratings were associated with higher scores on both SISQ constructs for the TD comparison group ($rs > .22, p < .05$), but not for those with WS. Also specific to TD participants, higher ratings on SISQ Social Approach were also associated with greater scores on MPQ Social Potency and Well-being ($rs > .33, p < .01$) subscales, as well as lower scores on MPQ Alienation ($r = -.26, p < .05$). In sum, a surprising finding showed that no factors indexing executive or cognitive ability, such as MPQ Control and MPQ Harm Avoidance, were associated with social behaviors as measured by the SISQ in either the WS or TD groups, suggesting the independence of social functions and

intelligence. Further, increased ambition and goal-driven behavior in individuals with WS was associated with a reduced tendency to approach others and initiate social interaction; by striking contrast, higher ambition and goal-orientation in TD was associated with increased initiation of interpersonal interactions. As expected, MPQ Social Closeness was most strongly associated with interpersonal behaviors as measured by the SISQ for both groups.

3.5. Regression Analysis: Personality Predictors of Social Functioning

Hierarchical linear regressions were then applied on the data to explore the predictive value of the MPQ Social Closeness subscale on the SISQ social behavior subscales. First, we controlled for chronological age and gender; second, we controlled for a set of personality traits that were observed to be associated with the SISQ subscales; and finally, our third block comprised the main personality trait of interest, Social Closeness (i.e., **Set 1:** age, gender, **Set 2:** Achievement, Alienation, Tradition, Well-being, Social Potency; **Set 3:** Social Closeness; refer to Table 3). Table 4 outlines the results of the linear regression analyses indicating the increased predictive value of MPQ Social Closeness above other traits and control variables for the WS group. In the regression analyses for predictors of Social Approach for WS, controlled demographic variables (age, gender) and personality factors had no significant predicting value; however, the inclusion of Social Closeness increased the percent of variance explained by 18.2%, ($F(8, 52) = 2.77, p < .05$). In this final model, Social Closeness emerged as the single most powerful predictor ($\beta = 0.59, p < .01$). Alternatively, the inclusion of Social Closeness did not significantly add explanatory power for the variance associated with the SISQ Social Approach behaviors for the TD group. Instead, the final significant model that accounted for the most variance in SISQ Social Approach solely included the block of control variables (age, gender) and the other MPQ personality traits excluding Social Closeness, ($F(7, 80) = 3.99, p < .001$). In sharp contrast to results for the WS group, Social Closeness did not significantly account for additional variance in SISQ Social Approach behavior for the TD group; rather, Age ($\beta = 0.31, p < .01$) and Social Potency ($\beta = 0.29, p < .05$) emerged as the best predictors.

Finally, Social Closeness accounted for an additional 16.7% of variance in SISQ Social Emotional behaviors for the WS group ($F(8, 52) = 6.14, p < .001$). The final model included all control variables and personality traits; however, Tradition ($\beta = 0.31, p < .05$) and Social Closeness ($\beta = 0.30, p < .001$) were the two strongest predictors of SISQ Social Emotionality for the participants with WS, while Social Closeness added no additional predictive value to the regression model with control variables and other traits in the TD participants. For the TD group, the final significant regression model included the Set 1 of control variables and Set 2 of MPQ personality traits ($F(7, 80) = 5.58, p < .001$). Of these predictors, Age ($\beta = -0.32, p < .01$) and Wellbeing ($\beta = 0.49, p < .01$) were the most powerful predictors of SISQ Social Emotionality. Taken together, while Social Closeness acts as a specific predictor that accounts for social behaviors beyond age, gender, and other personality traits as measured by the MPQ in individuals with WS, this is not the case for the TD individuals.

4. Discussion

The aim of the present exploratory study was to compare both the stability and the distinctiveness of personality traits as measured by the MPQ, in tandem with social-emotional tendencies as indexed by the SISQ, in participants with WS contrasted with age-matched TD comparison individuals. We specifically sought to elucidate how the differences in personality attributes between individuals with WS and TD may be related to their respective profiles of social functioning, which are currently poorly understood. This question is important, as improved understanding of how specific personality traits may be related to socially maladaptive behaviors over the course of development in WS may help develop intervention approaches aimed at improving social functioning of those with the syndrome.

The main finding revealed a robust difference in social motivation between individuals with WS and TD: whereas in individuals with WS, increased social-affiliative behaviors were driven by a desire for social closeness, i.e., predicted by the combination of gregarious, people-oriented, and affectionate personality features, for the TD group, social interactive behaviors were driven by a desire for social power, i.e., predicted by the combination of persuasive, dominant, and visible personality attributes. The result for the WS group is in good agreement with that of the previous study by Klein-Tasman and Mervis (2003), in that gregarious, people-oriented, tense, sensitive, and visible personality attributes distinguished their sample of individuals with WS from those with mixed developmental disability etiologies. However, as the study of Klein-Tasman and Mervis (2003) did not include a TD comparison group, and only included children aged 8-10 years, the current study extends the earlier findings in multiple ways. First, the current findings highlighted that the personality traits pertaining to social-affective qualities as indexed the MPQ appear to uniquely distinguish WS from TD individuals across a wide age range. Second, consistent with Klein-Tasman and Mervis (2003), the present results identified the personality characteristics cluster of Social Closeness as the main predictor of increased social-affiliative behavior in real life in WS. Finally, our study provides a novel contribution to current literature in WS by underscoring the major difference in the intimate nature of the social affiliative drive/motives in WS versus TD: interpersonal/affective versus social potency/dominance focus, respectively.

In terms of the between-group differences in personality traits as measured by the MPQ, relative to the WS group, the TD participants were rated significantly higher on the Achievement subscale, which taps into ambition, and tendency toward hard work and perseverance, at both developmental stages. By contrast, those with WS were rated significantly higher than the TD group on the personality factors assessing sociability and stress/anxiety. Whereas those with WS relative to TD were less reflective, neat, organized, and sensible as children, the between-group differences in this domain diminished with age. With regard to the stability of the personality traits within groups, no developmental differences were evident for either the Social Potency or the Social Closeness scores within either experimental group, suggesting that the respective traits characterizing the personality of individuals with WS and TD appear consistent across development. In both experimental groups, achievement-related traits (ambition, hard-working and persevering predisposition)

increased with age, whereas those relating to interpersonal and emotional aspects of personality (gregariousness, people-oriented and affectionate disposition) remained stable. Whereas those with WS remained stable on stress reaction (tense, sensitive, reverse of even-tempered), this personality dimension diminished with age in TD individuals. However, it is important to note that one limitation of the current study pertains to the cross-sectional design, which may mask some developmental effects. In light of the current pattern of findings, future research is warranted to explore such effects longitudinally.

We hypothesized that Social Closeness would act as a strong predictor of social behaviors in general; however, this was not the case for the TD group. Instead, Social Potency was the strongest determinant of social-affiliative behavior in typical individuals. These findings may implicate that in normative development, individuals typically form friendships and other relationships relatively more openly and frequently while young; thus, as individuals mature, the relationship between their age and sociability may gain in predictive value. Moreover, their affiliations may thus become more deliberate and measured with age, as they presumably have already formed much of their social networks by earlier life. In contrast, Social Closeness may act as a robust predictor of social behaviors in WS across lifespan due to their inability to form lasting friendships and other social relations. The poor social adjustment endorsed by those with WS (Gosch & Pankau, 1994) may therefore perpetuate the longing to integrate in social groups. Thus, one plausible interpretation of these results is that while individuals with WS continually socially engage with others in an attempt to build close relationships, TD individuals socially approach others mainly as a function of attention seeking together with their growing desire to demonstrate leadership and social-dominance, and not with the primary aim of forming friendships. It is noteworthy, however, that the low scores of individuals with WS on Social Potency may at least partially reflect the fact that such persons may have fewer opportunities for leadership than their TD counterparts; at the same time, leadership is only one aspect of the subscale. Nevertheless, the profile described herein for individuals with WS fits in well with Pober's recent notion that WS is associated with a "cocktail party personality" (Pober, 2010). Recently, a study by Klein-Tasman and colleagues (2011) suggested that despite the socially gregarious nature and eagerness to engage in social interaction with others, specifically problematic social behaviors pertaining to difficulties with perspective-taking and social communication contribute to the compromised social outcomes associated with WS.

Additionally, social emotionality was predicted by different personality factors in those with WS versus TD, with Social Closeness and Tradition as significant coefficients for those with WS. In contrast, for TD individuals, social emotional behaviors such as increased empathy were better predicted by increasing age and Well-Being, which indexes the degree they demonstrate positive thinking and affect. It is possible that in TD, those who are more positive in their cognitive and emotional styles may be more sensitive to others' emotions. Indeed, research on normative development suggests that positive and prosocial disposition is associated with increased empathy (Eisenberg & Miller, 1987; Eisenberg et al., 1996; Knafo et al., 2008). Correspondingly, many investigations have shown that increased negative affectivity and aggression are related to decreased empathy (Eisenberg et al., 1996; Miller & Eisenberg, 1988). Our evidence thus further contributes to this literature by indicating that as TD individuals mature, those with greater positive disposition in both

cognitive and emotional terms may be more likely to demonstrate greater social-affectivity, including increased empathic concern and emotional awareness of others. However, more normative developmental research is warranted to further elucidate the relationship between Well-Being and social emotional functioning, specifically in light of recent finding suggesting that different environmental influences significantly impact the continuity and stability of social affectivity, such as empathic concern (Knafo et al., 2008). Thus, it remains unclear whether the predictive effect of Well-Being and social affective functioning in TD individuals may be malleable in face of environmental influences.

In contrast, social emotionality in individuals with WS was best predicted by both Social Closeness and Tradition traits, regardless of age. A possible explanation for our results is that individuals with WS engage in social-affective interactions such as empathic gestures to conform to their perception of societal standards in efforts to fit in peer groups. It is possible that these individuals interpret their overly empathic tendencies and heightened attention towards others facial emotions as a „typical” behavior, and as such, engage in such social conduct in efforts to be part of social groups. However, considering that there are no known published studies investigating the link between social functioning and personality in individuals with WS, this phenomena should be further evaluated in larger samples across development in future studies.

Notably, individuals with WS were further consistently reported to show elevated stress and anxiety-related behaviors as compared to the TD individuals across development. Thus, consistent with the existing literature (Jabbi et al., 2012; Mervis & Klein-Tasman, 2000; Meyer-Lindenberg et al., 2006), the current study suggests that WS is associated with a unique personality characterized by overly social, emotional, and affectionate disposition coupled with increased stress and anxiety; traits that are relatively stable over time. The current findings deepen the existing characterization of the social phenotype of WS by adding ecological validity to previous reports: increased stress and anxiety indeed appear to coexist with increased social approachability in real life in WS. This is in line with previous reports suggesting that the anxiety associated with WS is specifically non-social in nature, relating to, e.g., anticipation about upcoming events, with a striking absence in relation to social contexts, e.g., interacting with unfamiliar people (Pober, 2010). This also combines with a failure to demonstrate separation anxiety as children upon being separated from their parents in WS (Mervis et al., 2012). Recently, a neuroimaging study by Jabbi et al. (2012) revealed that the neurogenetic basis of the hypersocial-anxious phenotype associated with WS implicates altered insula volume as well as aberrant connectivity between the insula and the amygdala and the OFC. In addition, the degree of these neurobiological alterations predicted the extent to which the participants with WS exhibited the characteristic personality profile specifically with respect to the MPQ personality attributes indexed mainly by Social Closeness.

One goal of the current study was to examine the potential contribution of personality traits other than those directly tapping into social-emotional attributes on social behaviors. Indeed, although one could have assumed a relationship between cognitive limitations and gregariousness in WS, our results suggest that increased social behaviors may instead stem from motivational aspects linked to personality, which in the case of WS likely reflect a

genetic basis (Jabbi et al., 2012), rather than, e.g., level of intelligence or a lack of impulse control. According to McGue, Bacon, and Lykken (1993), stable components of personality may be ascribed to “an enduring influence of genetic factors,” whereas changes in traits may be a result of environmental influences. Although entirely speculative, in the context of the higher Social Closeness in individuals with WS as compared to the TD comparison group, social motivation as measured by this personality dimension may potentially be linked to the genetic features underpinning WS. The current finding is of further relevance to the recent suggestion that the combination of intellectual disability and social dysfunction represents a particularly powerful risk factor of social vulnerability (Jawaid et al., 2011). The current results suggest that a more complicated mechanism underlies the overt social tendencies in both WS and TD. Indeed, cognitive and executive function traits related to behavioral inhibition, such as Control and Harm Avoidance, were surprisingly weakly correlated with social-affiliative behaviors in WS. As Control and Harm Avoidance factors index impulse control and planning, attributes that are also necessary for social-communicative skills, no association was found with either Social approach or Social-emotional subscales of the SISQ for either experimental group. Thus, although individuals with WS have been noted to place themselves at risk of harm by their indiscriminate approach behavior (Doyle et al., 2004), Harm Avoidance was dissociated with social-approach behaviors in the TD comparison group, suggesting that other mechanisms underlie normative social decision-making.

5. Conclusions

The present study extends the current literature by deepening the characterization of the nature of the social drive in WS by forging important links with the characteristic WS personality: whereas the motivation for increased social affiliation in individuals with WS appears to center around the desire to form interpersonal and affectionate relationships, TD individuals appear to be driven by social dominance. This reflects a major difference in the function of social overtures associated with WS and in TD. The current study highlighted similarities and differences in personality across different ages in individuals with WS and TD, providing additional support to the growing literature underscoring that the WS personality is best captured by increased sociability and anxiety; traits that are stable across development. In contrast, in normative development, social traits are subjected to change across development as these individuals transition from childhood to adolescence and finally adulthood, particularly with respect to increasing social confidence and friendship formation (Roberts, Caspi, & Moffitt, 2001). As Jawaid et al. (2011) note: “In the future studies, it is essential to bridge gaps between experimental observations and real-life experiences in WS” (p. 346) – indeed, our investigation made a step toward such a contribution by adding ecological validity to the previous speculations about the relationship between the WS personality and hypersociability, as a previous study has suggested that the SISQ provides a sensitive index of expressive social behavior in WS, unlike some receptive experimental measures (Järvinen-Pasley et al., 2010). Ultimately, the current study further demonstrates that future research directed at examining the distinct WS personality in combination with assessments indicative of ecologically valid social-affective behavior may offer a fruitful

avenue for future discoveries of therapeutic intervention approaches for enhancing social adjustment in those with WS.

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References

- Achenbach, T. Manual for the Child Behavior Checklist and Revised Child Behavior Profile. T. M. Achenbach; Burlington, VT: 1983.
- Achenbach, T. Structures of mood and personality and their relevance to assessing anxiety, with an emphasis on self-report.. In: Tuma, AH.; Maser, JS., editors. Anxiety and the anxiety disorders. Lawrence Erlbaum Associates, Inc.; Hillsdale, NJ: 1985. p. 681-716.
- Allen SR, Thorndike RM. Stability of the WAIS-R and WISC-III factor structure using cross-validation of covariance structures. *Journal of Clinical Psychology*. 1995; 51:648–657. [PubMed: 8801241]
- American Academy of Pediatrics, Committee on Genetics. Health care supervision for children with Williams syndrome (RE0034). American Academy of Pediatrics Policy Statement. 2001; 107:1192–1204.
- Bellugi U, Lichtenberger L, Jones W, Lai Z, St. George M. I. The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*. 2000; 12:7–29. [PubMed: 10953231]
- Canivez GL, Neitzel R, Martin BE. Construct validity of the Kaufman Intelligence Test, Wechsler Intelligence Scale for Children-Third Edition, and Adjustment Scales for Children and Adolescents. *Journal of Psychoeducational Assessment*. 2005; 23:15–34.
- Davies M, Udwin O, Howlin P. Adults with Williams syndrome. Preliminary study of social, emotional and behavioural difficulties. *British Journal of Psychiatry*. 1998; 172:273–276. [PubMed: 9614479]
- Doyle TF, Bellugi U, Korenberg JR, Graham J. “Everybody in the world is my friend” hypersociability in young children with Williams syndrome. *American Journal of Mental Retardation*. 2004; 124a:263–273.
- Dykens EM. Anxiety, fears, and phobias in persons with Williams syndrome. *Developmental Neuropsychology*. 2003; 23:291–316. [PubMed: 12730029]
- Eisenberg N, Fabes RA, Murphy B, Karbon M, Smith M, Maszk P. The relations of children's dispositional empathy-related responding to their emotionality, regulation and social functioning. *Developmental Psychology*. 1996; 32:195–209.
- Eisenberg N, Miller PA. The relation of empathy to prosocial and related behaviors. *Psychological Bulletin*. 1987; 101:91–119. [PubMed: 3562705]
- Gosch A, Pankau R. Social-emotional and behavioral adjustment in children with Williams-Beuren syndrome. *American Journal of Medical Genetics*. 1994; 53:335–339. [PubMed: 7864042]
- Gosch A, Pankau R. Personality characteristics and behavior problems in individuals with Williams syndrome. *Developmental Medicine and Child Neurology*. 1997; 39:527–533. [PubMed: 9295848]
- Jabbi M, Kippenhan JS, Kohn P, Marenco S, Mervis CB, Morris CA, Meyer-Lindenberg A, Berman KF. The Williams syndrome chromosome 7q11. 23 hemideletion confers hypersocial, anxious personality coupled with altered insula structure and function. *Proceedings of the National Academy of Sciences USA*. 2012; 109:E860–E866.
- Järvinen A, Korenberg JR, Bellugi U. The social phenotype of Williams syndrome. *Current Opinion in Neurobiology*. 2013; 23:414–422. [PubMed: 23332975]

- Järvinen-Pasley A, Adolphs R, Yam A, Hill KJ, Grichanik M, Reilly J, Mills D, Reiss AL, Korenberg JR, Bellugi U. Affiliative behavior in Williams syndrome: social perception and real-life social behavior. *Neuropsychologia*. 2010; 48:2110. [PubMed: 20385151]
- Järvinen-Pasley A, Bellugi U, Reilly J, Mills DL, Galaburda A, Reiss AL, Korenberg JR. Defining the social phenotype in Williams syndrome: A model for linking gene, the brain, and behavior. *Development and Psychopathology*. 2008; 20:1–35. [PubMed: 18211726]
- Javorsky J. The relationship between the Kaufman Brief Intelligence Test and the Wechsler Intelligence Scale for Children-III in a clinical sample. *Assessment for Effective Intervention*. 1993; 19:377–385.
- Jawaid A, Riby DM, Owens J, White SW, Tarar T, Schulz PE. 'Too withdrawn' or 'too friendly': Considering social vulnerability in two neuro-developmental disorders. *Journal of Intellectual Disability Research*. 2011; 56:335–350. [PubMed: 21801261]
- Jones W, Bellugi U, Lai Z, Chiles M, Reilly J, Lincoln A, Adolphs R. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*. 2000; 12:30–46. [PubMed: 10953232]
- Kaufman, AS.; Kaufman, NL. Kaufman Brief Intelligence Test manual. American Guidance Service; Circle Pines, MN: 1990.
- Klein-Tasman BP, Li-Barber KT, Magargee ET. Honing in on the social phenotype in Williams syndrome using multiple measures and multiple raters. *Journal of Autism and Developmental Disorders*. 2011; 41:341–351. [PubMed: 20614173]
- Klein-Tasman BP, Mervis CB. Distinctive personality characteristics of 8-, 9-, and 10-year-olds with Williams syndrome. *Developmental Neuropsychology*. 2003; 23:269–290. [PubMed: 12730028]
- Knafo A, Zahn-Waxler C, Van Hulle C, Robinson JL, Rhee SH. The developmental origins of a disposition toward empathy: Genetic and environmental contributions. *Emotion*. 2008; 8:737–752. [PubMed: 19102585]
- Korenberg JR, Chen XN, Hirota H, Bellugi U, Burian D, Roe B, Matsuoka R. Genome structure and cognitive map of Williams syndrome. *Journal of Cognitive Neuroscience*. 2000; 12:89–107. [PubMed: 10953236]
- Leyfer OT, Woodruff-Borden J, Klein-Tasman BP, Fricke JS, Mervis CB. Prevalence of psychiatric disorders in 4 to 16-year-olds with Williams syndrome. *American Journal of Medical Genetics Part B*. 2006; 141b:615–22.
- Martens MA, Hasinski AE, Andridge RR, Cunningham WA. Continuous cognitive dynamics of the evaluation of trustworthiness in Williams syndrome. *Frontiers in Psychology*. 2012; 3:160. [PubMed: 22675313]
- McGue M, Bacon S, Lykken D. Personality stability and change in early adulthood: A behavioral genetic analysis. *Developmental Psychology*. 1993; 29:96–109.
- Mervis CB, Dida J, Lam E, Crawford-Zelli NA, Young EJ, Henderson DR, Osborne LR. Duplication of GTF2I results in separation anxiety in mice and humans. *American Journal of Human Genetics*. 2012; 90:1064–1070. [PubMed: 22578324]
- Mervis CB, John AE. Cognitive and behavioral characteristics of children with Williams syndrome: Implications for intervention approaches. *American Journal of Medical Genetics Part B*. 2010; 154:229–248.
- Mervis CB, Klein-Tasman BP. Williams syndrome: cognition, personality, and adaptive behavior. *Mental Retardation and Developmental Disabilities Research Reviews*. 2000; 6:148–158. [PubMed: 10899809]
- Mervis CB, Robinson BF. Expressive vocabulary ability of toddlers with Williams syndrome or Down syndrome: A comparison. *Developmental Neuropsychology*. 2000; 17:111–126. [PubMed: 10916578]
- Meyer-Lindenberg A, Hariri AR, Munoz KE, Mervis CB, Mattay VS, Morris CA, Berman KF. Neural correlates of genetically abnormal social cognition in Williams syndrome. *Nature Neuroscience*. 2005; 8:991–993.
- Meyer-Lindenberg A, Mervis CB, Berman KF. Neural mechanisms in Williams syndrome: a unique window to genetic influences on cognition and behaviour. *Nature Reviews Neuroscience*. 2006; 7:380–393.

- Miller PA, Eisenberg N. The relation of empathy to aggressive and externalizing/antisocial behavior. *Psychological Bulletin*. 1988; 103:324–344. [PubMed: 3289071]
- Morris CA. The behavioral phenotype of Williams syndrome: A recognizable pattern of neurodevelopment. *American Journal of Medical Genetics Part*. 2010; C154:427–431.
- Muñoz KE, Meyer-Lindenberg A, Hariri AR, Mervis CB, Mattay VS, Morris CA, Berman KF. Abnormalities in neural processing of emotional stimuli in Williams syndrome vary according to social vs. non-social content. *Neuroimage*. 2010; 50:340–346. [PubMed: 20004252]
- Naglieri JA, Pfeiffer SI. Reliability and stability of the WISC-R for children with below average IQs. *Educational and Psychological Research*. 1983; 3:203–208.
- Ng R, Bellugi U. Physiological and psychological anxiety in adults with Williams syndrome, developmental delay, and typical development. *Neurology*. 2013; 80:P05.128.
- Pober BR. Williams–Beuren syndrome. *New England Journal of Medicine*. 2010; 362:239–252. [PubMed: 20089974]
- Prewett PN. The relationship between Kaufman Brief Intelligence Test (K-BIT) and WISC-R with referred students. *Psychology in the Schools*. 1992; 29:25–27.
- Reilly J, Losh M, Bellugi U, Wulfeck B. “Frog, where are you?” Narratives in children with specific language impairment, early focal brain injury, and Williams syndrome. *Brain and Language*. 2004; 88:229–247. [PubMed: 14965544]
- Riby DM, Hancock PJ. Viewing it differently: Social scene perception in Williams syndrome and autism. *Neuropsychologia*. 2008; 46:2855–2860. [PubMed: 18561959]
- Roberts BW, Caspi A, Moffitt TE. The kids are alright: Growth and stability in personality development from adolescence to adulthood. *Journal of Personality and Social Psychology*. 2001; 81:670–683. [PubMed: 11642353]
- Rothbart MK, Ahadi SA, Hershey KL, Fisher P. Investigations of temperament at three to seven years: The Children's Behavior Questionnaire. *Child Development*. 2003; 72:1394–1408. [PubMed: 11699677]
- Ryan JJ, Ward LC. Validity, reliability, and standard errors of measurement for two seven-subtest short forms of the Wechsler Adult Intelligence Scale – III. *Psychological Assessment*. 1999; 11:207.
- Sarimski K. Behavioural phenotypes and family stress in three mental retardation syndromes. *European Child and Adolescent Psychiatry*. 1997; 6:26–31. [PubMed: 9112044]
- Searcy YM, Lincoln AJ, Rose FE, Klima ES, Bavar N, Korenberg JR, Dykens EM. The relationship between age and IQ in adults with Williams Syndrome. *American Journal of Mental Retardation*. 2004; 108:231–236. [PubMed: 15072519]
- Spitz HH. Intratest and intertest reliability and stability of the WISC, WISC-R, and WAIS Full Scale IQs in a mentally retarded population. *The Journal of Special Education*. 1983; 17:69–80.
- Tellegen, A. Structures of mood and personality and their relevance to assessing anxiety, with an emphasis on self-report.. In: Tuma, AH.; Maser, D., editors. *Anxiety and the anxiety disorders*. Lawrence Erlbaum Associates, Inc.; Hillsdale, NJ: 1985. p. 681-716.
- Thornton-Wells TA, Avery SN, Blackford JU. Using novel control groups to dissect the amygdala's role in Williams syndrome. *Developmental Cognitive Neuroscience*. 2011; 1:295–304. [PubMed: 21731599]
- Tonge, BJ. Psychopathology of children with developmental disabilities.. In: Bouras, N., editor. *Psychiatric and behavioural disorders in developmental disabilities and mental retardation*. University Press; Cambridge, UK: 1999. p. 157-174.
- Udwin O. A survey of adults with Williams syndrome and idiopathic infantile hypercalcaemia. *Developmental Medicine and Child Neurology*. 1990; 32:129–141. [PubMed: 2338177]
- Udwin O, Yule W. A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*. 1991; 13:232–244. [PubMed: 1864913]
- Walters SO, Weavers KA. Relationships between the Kaufman Brief Intelligence Test and the Wechsler Adult Intelligence Scale-Third Edition. *Psychological Reports*. 2003; 92:1111–1115. [PubMed: 12931928]
- Wechsler, D. *Manual for the Wechsler Intelligence Scale for Children (rev.)*. The Psychological Corporation; San Antonio, TX: 1974.

- Wechsler, D. WAIS-R manual: Wechsler Adult Intelligence Scale (rev.). The Psychological Corporation; New York: 1981.
- Wechsler, D. Manual for the Wechsler Intelligence Scale for Children. 3rd ed.. The Psychological Corporation; San Antonio, TX: 1991.
- Wechsler, D. WAIS-III administration and scoring manual. 3rd ed.. The Psychological Corporation; San Antonio, TX: 1997.
- Wechsler, D. Wechsler abbreviated scale of intelligence. The Psychological Corporation; San Antonio, TX: 1999.
- Zitzer-Comfort C, Doyle T, Masataka N, Korenberg J, Bellugi U. Nature and nurture: Williams syndrome across cultures. *Developmental Science*. 2007; 10:755–762. [PubMed: 17973792]

Highlights

- Williams syndrome (WS) is characterized by a unique social personality profile
- Associations between personality traits and social functioning were examined
- The personality trait of Social Closeness was a robust predictor of social functioning in WS
- In typical development, the strongest predictor of interpersonal behaviors was Social Potency
- The personality attributes linked to interpersonal behaviors diverge in WS and TD

Table 1

Participant Characteristics.

	Williams Syndrome		Typical Development	
	< 18 years (<i>SD</i>)	Adults (<i>SD</i>)	< 18 years (<i>SD</i>)	Adults (<i>SD</i>)
N	28	27	34	53
Gender	15F	11F	17F	27F
Age (years)	10.64 (2.68)	33.41 (8.92)	9.84 (2.20)	29.46 (9.40)
VIQ	70.14 (14.85)	70.15 (9.59)	117.44 (11.10)	107.04 (11.09)
PIQ	67.82 (14.68)	66.56 (8.55)	106.09 (10.75)	104.34 (14.33)
FIQ	66.25 (14.16)	66.63 (9.52)	113.44 (10.14)	106.06 (11.17)

Table 2

Average Personality Trait Rating and Standard Deviation of Child/Adolescent and Adult Williams Syndrome and Typical Developing Participants (N=142).

Personality Factors	Williams Syndrome			Typically Developing		
	< 18 years (N=28)	Adults (N=27)	t-score	< 18 years (N=34)	Adults (N=53)	t-score
Absorption	2.90 (0.61)	2.89 (0.67)	0.09	2.64 (0.51)	2.78 (0.65)	-0.99
Achievement	1.70 (0.66)	2.17 (0.64)	-2.68**	2.63 (0.72)	3.16 (0.59)	-3.85**
Aggression	1.73 (0.79)	1.46 (0.53)	1.45	1.80 (0.66)	1.57 (0.51)	1.92 ⁺
Alienation	1.77 (0.64)	1.73 (0.68)	0.22	1.74 (0.61)	1.78 (0.72)	-0.40
Control	2.08 (0.61)	2.52 (0.53)	-3.11**	2.62 (0.63)	2.58 (0.68)	0.23
Harm Avoidance	3.00 (0.76)	2.74 (0.41)	1.56	2.75 (0.68)	2.56 (0.50)	1.13
Social Closeness	3.76 (0.38)	3.61 (0.54)	1.20	3.19 (0.66)	3.04 (0.77)	1.12
Social Potency	2.46 (0.55)	2.52 (0.56)	-0.36	2.76 (0.70)	2.75 (0.69)	0.19
Stress Reaction	2.76 (0.68)	2.47 (0.72)	1.55	2.32 (0.60)	1.98 (0.66)	2.14*
Traditionalism	2.75 (0.65)	3.09 (0.64)	-1.94 ⁺	2.98 (0.63)	2.99 (0.74)	-0.13
Well-Being	3.51 (0.51)	3.35 (0.60)	1.11	3.21 (0.69)	3.30 (0.58)	-0.60

*** $p < .001$

⁺ $p < .10$

* $p < .05$

** $p < .01$

Table 3

Pearson Correlations of the Multidimensional Personality Questionnaire and Salk Institute Sociability Questionnaire Subscales.

	Williams Syndrome (n=53)		Typical Development (n=81)	
	Social Emotionality	Social Approach	Social Emotionality	Social Approach
Absorption	.26	.20	.21	.07
Achievement	.13	-.22	.24*	.22*
Aggression	-.20	-.03	-.18	-.18
Alienation	-.15	-.09	-.15	-.26*
Control	.08	-.13	.06	-.12
Harm Avoidance	.24	.06	-.03	-.06
Social Closeness	.68***	.45***	.42***	.37***
Social Potency	.21	.15	.26*	.36***
Stress Reaction	.00	.01	-.04	-.18
Tradition	.45***	-.04	.31**	.11
Well-Being	.42**	.11	.49***	.33**

Table 4

Linear Regression Models: Personality Predictors of Salk Institute Sociability Questionnaire Subscales.

	Williams Syndrome (n=53)						Typical Development (n=81)					
	Social Emotionality			Social Approach			Social Emotionality			Social Approach		
	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)	B (SE)
Set 1												
Age	.00 (.01)	.00 (.01)	.00 (.01)	.01 (.01)	.01 (.01)	-.02 (.01)	-.02 (.01)	-.02 (.01)	.02 (.01)	.02 (.01)	.03 (.01)	.03 (.01)
Gender	-.02 (.28)	.11 (.24)	-.05 (.21)	.03 (.24)	.02 (.24)	.18 (.19)	.03 (.17)	-.05 (.18)	.17 (.20)	.13 (.19)	.01 (.19)	.01 (.19)
Set 2												
Achievement	.40 (.22)	-.13 (.20)		-.50 (.22)	-.26 (.21)		.01 (.16)	.02 (.16)		-.15 (.18)	-.13 (.18)	-.13 (.18)
Alienation	.03 (.25)	.09 (.22)		-.15 (.24)	-.25 (.22)		.16 (.14)	.22 (.15)		-.23 (.16)	-.14 (.16)	-.14 (.16)
Tradition	.77 (.23)*	.46 (.21)*		.10 (.22)	-.18 (.22)		.10 (.16)	.16 (.16)		-.15 (.17)	-.08 (.18)	-.08 (.18)
Well-Being	.56 (.31)	-.04 (.31)		.18 (.31)	-.36 (.32)		.67 (.20)*	.53 (.22)*		.32 (.22)	.14 (.24)	.14 (.24)
Social Potency	.31 (.26)	.09 (.24)		.42 (.26)	.22 (.24)		.05 (.14)	.00 (.14)		.38 (.15)**	.32 (.16)*	.32 (.16)*
Set 2												
Social Closeness			1.19 (.30)***		1.08 (.31)**			.24 (.16)				.32 (.17)
R ²	.06%	36.1%	52.75%	.05%	15.3%	33.5%	9.2%	34.9%	36.9%	8.7%	27.7%	31.0%
F-change	.02	5.08***	15.48***	.01	1.62	12.03***	3.96*	5.75***	2.34	3.70*	3.83***	3.52

 $p < .001$ **
 $p < .01$ *
 $p < .05$