Title: Relations of Executive Function and Intellectual Ability to Social Communication and Interaction and to Repetitive Behavior: Children with Williams Syndrome

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Introduction: Williams syndrome (WS) is caused by a hemizygous deletion of 26 – 28 genes on chromosome 7q11.23. A particularly notable characteristic of people with WS is their hypersociability, which manifests as an intense desire to approach and engage with others. At the same time, individuals with WS demonstrate considerable difficulty navigating social interactions, including trouble establishing and maintaining friendships. WS also is associated with difficulty with executive functioning, including skills related to behavioral and emotional regulation; these difficulties may play a role in the WS social and behavioral phenotype (Rhodes et al., 2010). For example, the social disinhibition shown by children with WS may relate to deficits inhibiting socially salient information (Porter et al., 2007). Moreover, the repetitive or restrictive behaviors that many children with WS engage in may be a consequence of problems disengaging or shifting attention (Lincoln et al., 2007). In this study we considered the contributions of the executive function domains of behavioral regulation and emotional regulation and of verbal and nonverbal intellectual ability to the social communication and interaction and repetitive behavior of children with WS.

Method: Participants were 141 children (77 girls) with genetically confirmed classic-length deletions of the WS region ranging in age from 6.00 to 17.98 years (*M* = 11.17 years). Parents completed the Social Responsiveness Scale – 2 (SRS-2; Constantino & Gruber, 2012), a parent-report measure of social impairment associated with autism spectrum disorder (ASD). The SRS-2 yields two DSM-5 compatible scales: Social Communication and Interaction (SCI) and Restricted Interests and Repetitive Behavior (RRB). Parents also completed the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000). Following the authors' recommendation, parents' responses to the BRIEF were scored using the BRIEF-2 norms (Gioia et al., 2015). The BRIEF-2 Behavior Regulation Index (BRI), which includes the Inhibit and Self-Monitor scales, and Emotional Regulation Index (ERI), which includes the Shift and Emotional Control scales, were used. The SRS-2 and BRIEF-2 yield *T*-scores (*M* = 50, *SD* = 10 for the general population); higher *T*-scores indicate greater difficulty. Participants completed the Kaufman Brief Intelligence Test – 2 (KBIT-2; Kaufman & Kaufman, 2004), a brief assessment of intellectual ability yielding separate verbal and nonverbal reasoning standard scores (SSs; *M* = 100, *SD* = 15).

Results: Pearson correlations were conducted to evaluate the relations of verbal ability and nonverbal reasoning ability to the measures of executive function and social responsiveness used in this study. KBIT-2 Verbal SS was significantly correlated with both SRS-2 RRB *T*-score (*r* = -.45, p < .001) and SCI *T*-score (*r* = -.32, *p* < .001), and KBIT-2 Nonverbal SS was significantly correlated with RRB *T*-score (*r* = -.32, p < .001). In contrast, Verbal SS and Nonverbal SS did not correlate significantly with BRIEF-2 BRI or ERI. Two multiple regression models were computed using the SRS-2 SCI and RRB *T*-scores as respective dependent variables. The independent variables included in both models were KBIT-2 Verbal SS, KBIT-2 Nonverbal SS, BRIEF2 BRI *T*-score, BRIEF2 ERI *T*-score, and sex. The first model statistically significantly predicted SRS-2 SCI *T*-score, F(5, 135) = 19.62, *p* < .001, adj. R^2 = .40, with KBIT-2 Verbal SS (*p* = .001, *sr* = -.23, BRIEF2 BRI *T*-score (*p* = .001, *sr* = .21), and BRIEF2 ERI *T*-score (*p* < .001, *sr* = .28) each accounting for a significant amount of unique variance. Likewise, the second model statistically significantly predicted SRS-2 RRB *T*-score, F(5, 135) = 36.63, *p* < .001, adj. R^2 = .55, with KBIT-2 Verbal SS (*p* < .001, *sr* = -.24), BRIEF2 BRI *T*-score (*p* = .038, *sr* = .12), and BRIEF2 ERI *T*-score (*p* < .001, *sr* = .40), each accounting for a significant amount of unique variance.

Discussion: SRS-2 RRB was significantly correlated with both verbal and nonverbal ability, and SRS-2 SCI was significantly correlated with verbal ability. This finding is consistent with the SRS-2's authors' (2012) report that SRS-2 Total T-score may be related to intelligence for children with intellectual disability. This lack of independence should be considered when interpreting the SRS-2 scores of children with WS. Results from the regression analyses revealed that the same three predictors (Verbal SS, BRI, and ERI) contributed significantly to both models. In relation to the total variance accounted for in the SCI model, the amount of unique variance accounted for by Verbal SS (5.34%), BRI (4.49%), and ERI (7.90%) was similar. In contrast, in relation to the total variance accounted for in the RRB model, the unique variance accounted for by ERI (15.76%) was notably greater

than that accounted for by Verbal SS (5.62%) or BRI (1.42%). The finding that verbal proficiency and behavioral and emotional regulation skills impact a wide breadth of social and behavioral outcomes highlights the importance of both language and behavioral and emotional regulation as intervention targets for children with WS.

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