2025 Gatlinburg Conference Poster Submission

Title: Characterizing Socio-communication in Children with Williams Syndrome Using the Social Responsiveness Scale-2

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Introduction: Children with Williams syndrome (WS) are often characterized as being gregarious and socially disinhibited (Kozel et al., 2021), with most children exhibiting high social approach to strangers. At the same time, in many ways the socio-communication phenotype associated with WS overlaps the socio-communication phenotype associated with autism spectrum disorder (ASD; Klein-Tasman et al., 2018). To further address this overlap, several studies have used the Social Responsiveness Scale-2 (SRS-2; Constantino & Gruber, 2012) to characterize the socio-communication skills of individuals with WS and the potential overlap with socio-communication skills in ASD. The two large-sample studies of individuals with WS that used the SRS-2 (Kopp et al., 2018; Hirari et al., 2022) included participants across a very broad age range (early childhood – middle adulthood) and did not report separate findings as a function of age. In the present study, we included more than twice as many participants and restricted the age range to childhood and adolescence, with the goal of providing a more precise description of the socio-communication phenotype of children with WS as measured by the SRS-2.

Method: Participants were 203 children (*M* = 8.91 years, *SD* = 4.37, range: 4 – 17.98; 103 females) with genetically-confirmed classic-length WS deletions. As part of a larger study, parents completed the SRS-2, a 65-item questionnaire that measures symptoms associated with ASD and yields sex-adjusted T-scores (*M* = 50, *SD* = 10 for the general population; higher T-scores indicate greater difficulty), which also are used to classify level of impairment. The SRS-2 includes five treatment subscales: Social Awareness, Social Cognition, Social Communication, Social Motivation, and Restricted Interests and Repetitive Behavior (RRB). Participants completed the Differential Ability Scales-2 (DAS-II; Elliot, 2007), a standardized assessment of intellectual abilities. Mean DAS-II General Conceptual Ability (GCA; similar to IQ) was in the mild intellectual disability range (*M* = 61.19, *SD* = 13.99, range: 30-94).

Results: Descriptive statistics are reported in Table 1 and proportions of individuals who scored at each severity level are reported in Table 2. SRS-2 T-scores were used in all statistical analyses. A repeated measures ANOVA, with Greenhouse-Geisser correction, indicated a significant effect of treatment subscale, $F(3.6, 722.2) = 201.9$, $p < .001$, $\eta_p^2 = .501$ and a significant interaction between treatment subscale and sex, $F(3.6, 722.2) = 4.16$, $p = .004$, $\eta_p^2 = .020$. The main effect of sex was not statistically significant, $F(1, 201) = .873$, $p = .351$, $\eta_p^2 = .004$. For treatment subscales, follow-up pairwise comparisons using Sidak correction indicated four distinct levels of skills: 1) Average performance on Social Motivation was in the typical range for the SRS-2 norming sample and significantly better than on any other treatment subscale (*d*s ≥ 1.12). 2) Average performance on Social Awareness and Social Communication was in the mild impairment range and did not differ significantly (*d* = .06). Performance on both subscales was significantly better than on RRB or Social Cognition (*d*s ≥ .25). 3) Average performance on RRB was in the moderate impairment range and significantly better than performance on Social Cognition (*d* = .25). 4) Average performance on Social Cognition also was in the moderate impairment range and significantly worse than on all other treatment subscales. Findings from follow-up pairwise comparisons using Sidak correction to address the significant interaction between treatment scale and sex indicated that the only significant difference was for Social Awareness. For this subscale, performance was significantly worse ($p = .009$, $d = .37$) for males (*M* = 66.73, *SD* = 9.28) than females (*M* = 63.06, *SD* = 10.61). Pearson correlations between GCA and treatment subscale T-score, although statistically significant, were small (-.22) to moderate (-.35). Correlations between chronological age and treatment subscale T-score were very small (-.14 to .06).

Table 1. Descriptive Statistics for SRS-2 T-scores (N = 203)

	M	*SD*	Median	Range
Treatment Subscales				
Social Awareness	64.87	10.12	65.00	35-95
Social Cognition	69.94	9.59	70.00	41-92
Social Communication	64.37	9.16	64.00	42-90
Social Motivation	52.43	8.89	51.00	38-89
RRB	67.55	11.63	68.00	42-96

Note. RRB = Restricted Interests and Repetitive Behavior.

Table 2. Proportion of Participants at Each SRS-2 Severity Level

	Typical	Mild	Moderate	Severe
Treatment Subscales				
Social Awareness	30.54%	24.14%	32.02%	13.30%
Social Cognition	14.29%	18.72%	35.96%	31.03%
Social Communication	30.54%	26.11%	30.54%	12.81%
Social Motivation	82.26%	8.87%	7.39%	1.48%
RRB	24.14%	21.18%	28.57%	26.11%

Note. RRB = Restricted Interests and Repetitive Behavior.

Discussion: These results highlight the considerable overlap between the socio-communication phenotypes of WS and ASD, which is consistent with previous findings (Hirai et al., 2022; Klein-Tasman et al., 2018; Kopp et al., 2018). Although GCA was significantly correlated with performance on the treatment subscales, the correlations were relatively small so accounted for little of the variance. The large sample size over a considerably more limited age range than previous studies allowed us to more precisely describe the WS social communication profile as measured by the SRS-2. Findings indicated four distinct levels of performance as a function of treatment scale, from typical for the general population to moderate/severe impairment: 1) Social Motivation, 2) Social Awareness and Social Communication, 3) RRB, 4) Social Cognition. This pattern provides further evidence that the social behavior of children with WS is consistent with Wing's description of the "active but unusual" sub-group of individuals with ASD. Future research with children with WS should consider how well individual children fit the group profile of socio-communicative abilities identified in this study and explore the longitudinal trajectories of their socio-communication skills.

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