Title: Executive Functions of Children and Adolescents with Williams Syndrome

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Introduction: Individuals with Williams syndrome (WS) often evidence difficulties in several aspects of executive functioning, including working memory, monitoring, inhibition, planning, and organization (Hocking, Reeve, & Porter, 2015). The aim of this study was to characterize the executive functioning of a large sample of children and adolescents with WS based on a widely used parent report measure, the Behavior Rating Inventory of Executive Function, second edition (BRIEF-2, Gioia, Isquith, Guy, & Kenworthy, 2015). Halvorsen and colleagues (2019) showed that relative to the first edition of the BRIEF (Gioia, Isquith, Guy, & Kenworthy, 2000), the BRIEF-2 three-factor model separating the inhibitory behavioral control dimension from the emotional control and metacognitive problem-solving dimensions had a better factor structure than the original two-factor BRIEF model and should be utilized for assessing the executive functioning of children with neurodevelopmental disorders.

Method: Participants were 306 children with WS (M = 11.23 years, SD = 3.64, 6 – 17.98 years, 156 girls). Parental report of executive function was measured by the BRIEF questionnaire, which includes all of the items that contribute to BRIEF-2 T-scores. Following the BRIEF-2 authors’ recommendations, the BRIEF questionnaires were scored based on the BRIEF-2 norms. The BRIEF-2 yields three indexes. The Behavior Regulation Index (BRI) includes the Inhibit and Self-Monitor scales. The Emotion Regulation Index (ERI) includes the Shift and Emotional Control scales. The Cognitive Regulation Index (CRI) includes the Initiate, Working Memory, Plan/Organize, Task-Monitor, and Organization of Materials scales. For the general population, all T-scores have a mean of 50 (SD = 10), with higher T-scores indicating greater difficulty. T-scores between 65 and 69 are considered potentially clinically elevated, and T-scores of 70 or higher are considered clinically elevated. 218 participants also completed the Differential Ability Scales-II (DAS-II; Elliott, 2007) from which General Conceptual Ability (GCA; similar to IQ) was calculated.

Results: Pearson correlations indicated weak relations between GCA (M = 61.98, SD = 13.96) and T-scores on the three indexes (BRI: r = -.16, p = .022; ERI: r = -.18, p = .007; CRI: r = -.002, p = .977) and between chronological age and the Index T-scores (BRI: r = -.25, p < .001; ERI: r = -.17, p = .170; CRI: r = .10, p = .099). A repeated measures ANOVA indicated a significant difference as a function of Index, F(2,610) = 45.10, p < .001, η² = .13. Performance on the BRI (M = 62.97, SD = 10.39) was significantly better than on the ERI (M = 64.42, SD = 10.13), which was significantly better than on the CRI (M = 67.86, SD = 8.29) (ps ≤ .012). 42% of the children scored in the potentially clinically elevated or clinically elevated range (T-scores ≥ 65) on the BRI, 56.4% on the ERI, and 63.1% on the CRI. Within the BRI, performance was significantly better on the Inhibit scale (M = 61.32, SD = 11.07) than on the Self-Monitor scale (M = 62.76, SD = 10.07), t(305) = -2.54, p = .011, d = .14. Within the ERI, performance for Emotional Control (M = 62.17, SD = 10.21) was significantly better than for Shift (M = 64.87, SD = 10.85), t(305) = 4.74, p < .001, d = .28. A repeated measures ANOVA indicated a significant difference as a function of CRI Scale, F(3.74, 1140.18) = 119.98, p < .001, η² = .28. Performance was significantly better on Organization of Materials (M = 59.44, SD = 9.05) than on the other scales (ps < .001). Performance on Plan/Organize (M = 64.81, SD = 8.79) was significantly better than on Initiate (M = 67.88, SD = 9.02), Task-Monitor (M = 67.88, SD = 8.78), and Working Memory (M = 68.31, SD = 8.57) (ps < .001). Performance on the latter three scales did not differ significantly (ps ≥ .98).
Discussion: These results highlight the presence of executive functioning difficulties in a large sample of children and adolescents with WS based on parent report. The initiation and task-monitoring problems identified likely contribute to the difficulties with mastery motivation previously found for young children with WS, whose mastery motivation was significantly weaker than that of same-aged children with Down syndrome even though the latter had more limited cognitive abilities (Rowe, 2007). These results highlight the importance of designing and delivering research-based interventions to improve execution functioning.

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