

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 Functioning, 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, items from the BRIEF were converted and scored based on the BRIEF-2 norms. The BRIEF-2 yields three indexes. The Behavior Regulation Index (BRI) includes Inhibit and Self-Monitor scales. The Emotion Regulation Index (ERI) includes Shift and Emotional Control. The Cognitive Regulation Index (CRI) includes Initiate, Working Memory, Plan/Organize, Task-Monitor, and Organization of Materials. For the general population, all T-scores have a mean of 50 ($SD = 10$), and T-scores between 65 and 69 are considered potentially clinically elevated, and at or above 70 clinically elevated (higher T-scores indicate greater difficulty). 218 participants ($M = 11.15$ years, $SD = 3.83$, 6 – 17.98 years, 110 girls) also completed the Differential Ability Scales-II (DAS-II; Elliott, 2007) and General Conceptual Ability (GCA, similar to IQ) SS was calculated. For the general population, SS have a mean of 100 ($SD = 15$).

Results: Pearson correlations indicated weak relations between GCA ($M = 61.98$, $SD = 13.96$, 28 – 94) and the three indexes (BRI: $r = -.16$, $p = .022$; ERI: $r = -.18$, $p = .007$; CRI: $r = -.002$, $p = .977$) and between chronological age and the three indexes (BRI: $r = -.25$, $p < .001$; ERI: $r = -.17$, $p = .170$; CRI: $r = .10$, $p = .099$). Repeated measures ANOVA was performed comparing the 3 indexes, $F(2,610) = 45.10$, $p < .001$, $\eta^2 = .13$. Performance on BRI ($M = 62.97$, $SD = 10.39$, 39 - 90) was significantly better than ERI ($M = 64.42$, $SD = 10.13$, 39 - 90), which was significantly better than CRI ($M = 67.86$, $SD = 8.29$, 40 - 87) ($ps \leq .012$). 42% percent of the children scored in either the potentially clinically elevated or clinically elevated ranges (T-scores ≥ 65) on the BRI, 56.4% on the ERI, and 63.1% on the CRI. Within BRI, performance was significantly better on the Inhibit scale ($M = 61.32$, $SD = 11.07$, 39 - 90) than on the Self-Monitor scale ($M = 62.76$, $SD = 10.07$, 39 - 90), $t(305) = -2.54$, $p = .011$, $d = .14$. Within ERI, Emotional Control ($M = 62.17$, $SD = 10.21$, 40 - 84) was significantly better than Shift ($M = 64.87$, $SD = 10.85$, 40 - 90), $t(305) = 4.74$, $p < .001$, $d = .28$. Repeated measures ANOVA was performed comparing the five scales of the CRI, $F(3.74, 1140.18) = 119.98$, $p < .001$, $\eta^2 = .28$. Performance was significantly better on Organization of Materials ($M = 59.44$, $SD = 9.05$, 38 - 81) than in any of the other four scales ($ps < .001$). Plan/Organize ($M = 64.81$, $SD = 8.79$, 37, 80) was significantly better than Initiate ($M = 67.88$, $SD = 9.02$, 38 - 84), Task-Monitor ($M = 67.88$, $SD = 8.78$, 35 - 82), and Working Memory ($M = 68.31$, $SD = 8.57$, 45 - 87) ($ps < .001$), and performance on these three scales did not differ ($ps \geq .98$). Within CRI, 60.8% performed at

or above 65 on Initiate, 67.3% on Task-Monitor, and on 67.3% Working Memory.

Discussion: These results highlight the presence of executive functioning difficulties in a large sample of children and adolescents with WS based on parent report. More difficulties were found for Initiate, Task-Monitor, and Working Memory. Initiate and task-monitor problems are likely to contribute to the mastery motivation problems that were previously found to be even more pervasive in young children with WS when compared to children with Down syndrome with more limited cognitive abilities (Rowe, 2007). These results highlight the importance of developing and providing interventions to prevent and improve these abilities. More studies that aim to understand the relation of executive functioning with adaptive behaviors and other cognitive abilities are necessary. Assessing the stability of these constructs is also an important area of research. Theoretical and practical implications will be discussed.

References:

- Elliott, C. D. (2007). *Differential ability scales, Second edition*. San Antonio, Texas. Psychological Corporation.
- Gioia, G. A., Isquith, P. K., Guy, S. C., & Kenworthy, L. (2000). *Behavior Rating Inventory of Executive Function (BRIEF)*. Lutz, FL: Psychological Assessment resources, Inc.
- Gioia, G. A., Isquith, P. K., Guy, S. C., & Kenworthy, L. (2015). *BRIEF-2: Behavior Rating Inventory of Executive Function: Professional Manual*. Psychological Assessment Resources.
- Halvorsen, M., Mathiassen, B., Amundsen, T., Ellingsen, J., Brøndbo, P. H., Sundby, J., ... & Martinussen, M. (2019). Confirmatory factor analysis of the behavior rating inventory of executive function in a neuro-pediatric sample and its application to mental disorders. *Child Neuropsychology, 25*, 599-616.
- Hocking, D. R., Reeve, J., & Porter, M. A. (2015). Characterising the profile of everyday executive functioning and relation to IQ in adults with Williams syndrome: Is the BRIEF adult version a valid rating scale?. *PLoS one, 10*(9), e0137628.
- Rowe, M. L. (2007). *Mastery motivation in young children with Williams syndrome or Down syndrome (Doctoral dissertation)*. Retrieved from ProQuest.