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Reliability of informant-report measures of executive functioning in children with Down syndrome

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Abstract

The current study evaluates the psychometric properties of the Behavior Rating Inventory of Executive Function (BRIEF) with children with Down syndrome. Caregivers of 84 children with Down syndrome rated their child's behavior with the BRIEF. Teacher-ratings were obtained for 57 children. About 40% of children with Down syndrome were reported by parents, and 70% by teachers, to exhibit clinically significant challenges with executive functioning. Distribution of scores was normal, internal consistency for subscales was questionable to primarily excellent, and inter-rater reliability was poor to good. Normative data conversions controlled for age, IQ and gender differences, with some exceptions. The study findings suggest that the BRIEF and its subscales generally performed in a psychometrically sound manner among children with Down syndrome.

Keywords

Down syndrome; trisomy 21; executive functioning; measurement; children

Individuals with Down syndrome often present with a distinct pattern of behavioral and cognitive skills (Dykens, Hodapp, & Finucane, 2000). Within this distinct behavioral and cognitive phenotype, there is variability across individuals in different domains, including self-regulation, social-emotional functioning, cognition, attention, language and motor skills (Silverman, 2007). Several of these areas of impairment fall under the category of executive functioning. Specifically relative to their typically developing peers, individuals with Down syndrome exhibit challenges in inhibitory control, in their ability to set-shift, and with working memory (Daunhauer et al., 2014; Lee et al., 2011). These deficits can have a substantial impact on daily functioning in the child with Down syndrome. Inattention and

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behavior regulation problems have been associated with decreased motivation and compliance in early childhood, and such deficits may impede adherence to early intervention (Gilmore & Cuskelly, 2009). Further, clinically significant problems with attention and behavior regulation independently and significantly predict deficits in adaptive skills in adolescents with Down syndrome (Jacola, Hickey, Howe, Esbensen, & Shear, 2014). The impact of these deficits in executive functioning for individuals with Down syndrome underscore the importance of better understanding these constructs with this population.

Research on executive functioning in individuals with Down syndrome, specifically the core areas of weakness in inhibitory control, the ability to shift, and working memory, has more commonly focused on neuropsychological or laboratory assessment of skills (for review see Lee et al., 2011). Inhibitory control is defined as the ability to curb or regulate attentional or behavioral responses. Findings of a relative weakness in inhibitory control among children with Down syndrome on neuropsychological assessments are inconsistent, with evidence for and also not replicating this finding (Borella, Carretti, & Lanfranchi, 2013; Carney, Brown, & Henry, 2013; Costanzo et al., 2013; Lanfranchi, Jerman, Dal Pont, Alberti, & Vianello, 2010; Pennington, Moon, Edgin, Stedron, & Nadel, 2003). Shifting is defined as the ability to transition from one task or activity to another, including the ability to demonstrate cognitive flexibility and to switch between tasks. Neuropsychological assessments corroborate a relative weakness in shifting for children with Down syndrome (Campbell et al., 2013; Carney, Brown, et al., 2013; Costanzo et al., 2013; Landry, Russo, Dawkins, Zelazo, & Burack, 2012; Lanfranchi et al., 2010). Working memory is a limited capacity system used to store and manipulate information in support of problem solving and goal-directed behavior (Baddeley, 1998). Working memory accounts for a substantial portion of developmental variance in global intelligence in typically developing children (Fry & Hale, 1996). Neuropsychological assessment demonstrates difficulties with working memory among children with Down syndrome, with relative weaknesses in verbal working memory as compared to visual working memory (Baddeley & Jarrold, 2007; Carney, Henry, et al., 2013; Costanzo et al., 2013; Landry et al., 2012; Lanfranchi et al., 2010; Rowe, Lavender, & Turk, 2006). However, others have demonstrated that children with Down syndrome present with comparable performance on working memory tasks relative to typically developing children matched for mental age and instead present with a specific difficulty with long-term memory tasks (Pennington et al., 2003).

More recent efforts have expanded assessment of executive functioning to include parent- and teacher-reports (Lee et al., 2011). Similar to the inconsistent support for deficits in inhibitory control when assessed with neuropsychology assessments, inconsistent findings are reported between parent- and teacher-reports of inhibitory control. Current findings support a relative weakness in inhibitory control when assessed with parent-reports but not with teacher-reports of executive functioning (Daunhauer et al., 2014). In contrast to the relative weakness in the ability to shift found on neuropsychology assessments, neither parent- nor teacher-reports of executive functioning demonstrate an area of weakness in the ability to shift relative to the child's mental age (Daunhauer et al., 2014). Yet parents and teachers demonstrate agreement in identifying challenges with working memory in children with Down syndrome (Daunhauer et al., 2014). These patterns of agreements and differences in executive functioning may reflect similar or different expectations across environments,

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where there are differences in the structure of tasks, the number of task demands, the number of individuals present, and the difficulty of tasks.

Given the developmental level of individuals with Down syndrome, the above mentioned use of informant-reports of executive functioning has focused on evaluating levels of executive functioning relative to the child's mental age (Lee et al., 2011; Pritchard, Kalback, McCurdy, & Capone, 2015). When accounting for mental age, parent-ratings on the Behavior Rating Inventory of Executive Function – Preschool version (BRIEF–P) demonstrate a pattern of deficits in working memory and planning, but not in inhibition or emotional control among young children with Down syndrome (Daunhauer et al., 2014; Gioia, Espy, & Isquith, 2003; Lee et al., 2011). However, this approach requires scoring a child on the BRIEF-P based on their mental age and not on their chronological age. Thus, items may not be age-appropriate to the child's current environment (gets out of control more than *playmates*; has trouble finishing tasks [such as games, puzzles, *pretend play activities*]). Further, methods for calculating mental age vary drastically depending on what measure of cognition or adaptive behavior is used, which can result in floor effects when used with adults with Down syndrome (D'Ardhuy et al., 2015).

There is a significant need for reliable and valid outcome measures for use in clinical trials in patients with Down syndrome in order to detect changes in response to treatment or intervention. Valid measures of executive functioning are especially needed as these skills have been the target of several clinical trials (D'Ardhuy et al., 2015; Kishnani et al., 2010).

This need is highlighted by the United States National Institutes of Health (NIH) convening Down syndrome Working Groups to evaluate clinical outcome measures for use with individuals with Down syndrome, including measures of executive functioning (NICHD, 2015). The BRIEF-P and BRIEF were both recommended by the NIH Down syndrome Working Group as appropriate for use with individuals with intellectual and development disabilities; however, further re-evaluation was recommended with regard to the use of these instruments in Down syndrome specifically (Esbensen et al., 2017). Currently, the BRIEF-P demonstrates moderate convergent validity with laboratory assessments of working memory, and adequate test-retest reliability when administered to parents/caregivers of adolescents and adults with Down syndrome and scores created based on their mental age (D'Ardhuy et al., 2015). Further, exploratory factor analysis of the BRIEF-P with children with Down syndrome ages 3–13 years supports the original three composite indices (Pritchard et al., 2015). Similarly, the BRIEF demonstrates good convergent validity with some laboratory assessments of inhibitory control and working memory in children with Down syndrome, but with some findings not replicating in other studies (Edgin et al., 2010; Esbensen & Hoffman, 2017). Interrater agreement has not been evaluated for all subscales of the BRIEF with children with Down syndrome, but preliminary evidence identifies good interrater reliability for the Inhibit and Working Memory subscales (Esbensen & Hoffman, 2017). The NIH Down syndrome Working Groups identified the paucity of empirical evidence for using currently available parent-report measures of executive functioning with individuals with Down syndrome (Esbensen et al., 2017; NICHD, 2015).

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The BRIEF is a measure worth evaluating based on the preliminary evidence of psychometric properties when used with children with Down syndrome. Although others have presented a good rationale for using the BRIEF-P scored on the mental age of children with Down syndrome, evaluation of the BRIEF based on the child's chronological age is warranted prior to any adaptations being made to scoring of the BRIEF-P. The BRIEF is also a worthy target of investigation as it includes age- and gender-based norms and clinical cut-offs identifying areas of concern. Preliminary findings with small sample sizes suggest that some BRIEF subscales, specifically Shift, may vary based on gender (Maiman et al., 2017). Similarly, this preliminary study also suggested that scores on the BRIEF may be impacted by the level of cognitive ability of the child.

Given the concern for challenges with executive functioning, as well as the lack of empirically supported parent- and teacher-report measures of executive functioning in children with Down syndrome, this study aimed to evaluate the psychometric properties of the BRIEF for use with children with Down syndrome. First, we described the rates and distribution of different problems with executive functioning at the subscale and index level to understand the pattern of behavior of concern from parent- and teacher-reports. Second, we examined the internal consistency of the BRIEF subscale and index scores from parent- and teacher-reports. Third, we examined the inter-rater reliability of subscale and index scores on the parent- and teacher-reports of the BRIEF. Last, we examined age, gender, and IQ differences on the BRIEF subscale and index scores after conversion to t-scores with normative data. Understanding the psychometric properties of the BRIEF with children with Down syndrome will inform the use of this measure and support the identification of reliable measures when reporting on problems with executive functioning. Identifying reliable and valid measures of executive functioning are needed to detect true change in this skill during clinical and behavioral interventions and during clinical trials.

Method

Participants

Parents of 84 children with Down syndrome completed rating forms as part of several larger clinical and community-based studies on behavior and cognition (Figure 1). Children with Down syndrome were between 6 to 18 years of age (M = 11.36 years, SD = 3.00), were primarily male (60.7%) and Caucasian (84.5%). Standard scores were obtained on different measures of cognition in this retrospective study, depending on the clinical or research purpose of the prior clinical or community study. Standard IQ scores are summarized in Table 1 by cognitive test. Ratings of adaptive behavior on the Scales of Independent Behavior – Revised were available on approximately half of the children (M = 48.5, SD = 20.8, range 0–93). Respondents were primarily mothers (93%), with fathers completing 3.5% of forms and both parents working together to complete 3.5% of forms.

Procedure

Families were recruited based on the age of the child and a diagnosis of Down syndrome. Families were recruited through a pediatric medical center, a Down syndrome specialty clinic, and through newsletters distributed by the local Down syndrome association. Clinical

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chart review contributed 36 children to the current sample where parent- and teacher-ratings of executive functioning were documented (see Figure 1). The remaining 48 children were recruited from community studies on Down syndrome. In community studies, parents provided information on the child's demographics and completed rating scales of the child's executive functioning on the BRIEF. Within a week, teachers also completed rating forms on the child's executive functioning on the BRIEF-teacher form that were distributed and collected through parents. Teacher-reports were collected from 57 teachers across community studies and clinical records. Teacher-reports were not documented in 13 clinical charts, were not obtained for 12 children in community-recruited research studies as the child was on school breaks, and was not obtained as part of the study protocol in one research study involving 2 children. All study activities were approved and overseen by the Institutional Review Board at the medical center.

Measures

The BRIEF (5–18) Parent and Teacher Forms are rating scales of everyday skills measuring executive functioning (Gioia, 2000). The BRIEF includes 86 items that measure skills of inhibition, shifting attention, emotional control, initiating tasks, problem-solving, working memory, organization and monitoring activities. Items are rated on a 3-point Likert-type scale from (1) Never to (3) Often. The BRIEF provides both subscale scores on the skills mentioned, and index scores of Behavioral Regulation Index (BRI), Metacognitive Index (MI) and a Global Executive Composite (GEC). The BRIEF subscales of inhibition and working memory demonstrate good convergent validity with hippocampal and prefrontal neuropsychology assessments when used with children with Down syndrome (Edgin et al., 2010).

Data Analysis

For the purpose of the current analyses, age and gender-standardized mean t-scores were calculated for the BRIEF parent- and teacher-report subscale and index scores. The percentage of children scoring above the clinical range of concern was calculated. The clinical range of concern was considered a t-score above 65, equating to 1.5 standard deviations outside of normative expectations (Gioia, 2000). The distribution of item and subscale scores was assessed and concerns with skew or kurtosis identified. Skew was considered a concern if the statistic was less than −0.8 or greater than 0.8. Kurtosis was considered a concern if the statistic was less than −3.0 or greater than 3.0.

Internal consistency of each BRIEF parent- and teacher-report subscale and index score was calculated using Cronbach's alpha. Interrater-reliability was calculated using the two-way mixed average measures form of intra-class correlation coefficients (ICC) comparing subscale and index scores on the parent- and teacher-forms of the BRIEF.

To assess for possible gender differences on subscale scores, two-sample t-tests were conducted on the BRIEF parent- and teacher-report subscale and index t-scores. Frequency comparisons (i.e., Chi-Square tests) were conducted to compare the frequency of clinically elevated scores by gender on BRIEF parent- and teacher-report subscale and index scores.

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Correlations were run to assess for possible associations with age on the BRIEF parent- and teacher-report t-scores.

As different measures of cognition were used, a median-split was created to dichotomize IQ and reduce potential variability across measures. T-tests were used again to assess for differences in BRIEF parent- and teacher-report t-scores for children with IQ scores above or below the median IQ score of 42.

Results

Group differences were run to assess comparability between community and clinical samples. Groups did not differ on age ($t(82) = 1.58$, $p = .12$), gender ($\chi(1)^2 = .01$, $p = .91$), race ($\chi(3)^2 = 3.43$, $p = .33$) or IQ (median split; $\chi(1)^2 = 1.43$, $p = .23$). Compared to those in the community-based samples, children seen in clinic were reported to have higher t-scores on parent-report subscale of Emotional Control (clinic $M = 57.0$, $SD = 13.0$; community $M = 51.3$, $SD = 10.8$; $t(82) = 2.19$, $p = .03$), and on the teacher-report subscales of Emotional Control (clinic $M = 72.6$, $SD = 17.8$; community $M = 62.3$, $SD = 14.6$; $t(55) = 2.39$, $p = .02$), and Monitor (clinic $M = 78.4$, $SD = 12.7$; community $M = 68.7$, $SD = 14.3$; $t(67) = 2.58$, $p = .01$).

Frequency of Problems with Executive Functioning

Table 2 presents the means, standard deviations and range of t-scores for BRIEF parent- and teacher-report subscale and index scores and the rates at which children met clinical criteria (frequency of scores ≥ 1.5 standard deviations above the mean). On the BRIEF parent-report, over 40% of children with Down syndrome in the sample met the criteria for clinical concern for the Global Executive Composite. On the BRIEF parent-report subscales, between 9–56% of children met the criteria for clinical concern on any subscale. The areas of executive functioning of most concern for children with Down syndrome were Monitor (56%) and Working Memory (51%). The areas of executive functioning of least concern were Organization of Materials (9%) and Emotional Control (16%). The distribution of BRIEF parent-report subscale and index scores demonstrated no concerns, demonstrating statistics for skew and kurtosis within the acceptable range.

On the BRIEF teacher-report, over 70% of children with Down syndrome in the sample met the criteria for clinical concern for the Global Executive Composite. A significantly greater frequency of children with Down syndrome were rated above the threshold on the Global Executive Composite on teacher ratings (71.7%) as compared to parent ratings (41.4%) ($\chi[1]^2 = 11.14$, $p < .001$). On the BRIEF teacher-report subscales, between 36–75% of children met the criteria for clinical concern on any subscale. The areas of executive functioning of most concern for children with Down syndrome were Working Memory (75%) and Initiate (70%). The areas of executive functioning of least concern were Organization of Materials (36%) and Plan/Organize (49%). The distribution of BRIEF teacher-report subscale and index scores demonstrated no concerns, demonstrating statistics for skew and kurtosis within the acceptable range.

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Inter-correlations between BRIEF subscale and index scores are presented in Table 3 (parent-report) and Table 4 (teacher-report). Scores on the BRIEF parent-report subscale and index scores did not significantly differ for children who had (n=57) or did not have (n=27) teacher-reports returned (Inhibit $t[82] = -0.26$, $p = .79$; Shift $t[82] = 0.33$, $p = .74$; Emotional Control $t[82] = 0.58$, $p = .56$; BRI $t[80] = 0.29$, $p = .78$; Initiate $t[80] = 0.85$, $p = .40$; Working Memory $t[82] = -0.22$, $p = .82$; Plan/Organize $t[68] = -0.13$, $p = .90$; Organization of Materials $t[80] = -0.09$, $p = .92$; Monitor $t[80] = -0.69$, $p = .49$; MI $t[66] = -0.33$, $p = .74$; GEC $t[68] = -0.17$, $p = .87$). Children for whom teacher-reports were obtained did not significantly differ from children with Down syndrome who did not have teacher-reports returned on age ($t[82] = 0.84$, $p = .33$), gender ($\chi[1]^2 = 1.31$, $p = .25$), race ($\chi[3]^2 = 2.44$, $p = .49$), or IQ ($\chi[1]^2 = 3.17$, $p = .08$)

Internal Consistency

Internal consistency for the BRIEF parent- and teacher-subscales are presented in Table 2. The alpha coefficients for the BRIEF parent-subscales ranged from .67 (questionable) to .96 (excellent). Only one subscale presented with questionable internal consistency (Initiate), one with acceptable internal consistency (Shift), six with good internal consistency (Inhibit, Emotional Control, Working Memory, Plan/Organize, Organization of Materials, and Monitor), and the three index scores all demonstrated excellent internal consistency (BRI, MI, GEC).

The alpha coefficients for the BRIEF teacher-subscales ranged from .76 (acceptable) to .97 (excellent). Only one subscale presented with acceptable internal consistency (Organization of Materials), five with good internal consistency (Shift, Initiate, Working Memory, Plan/Organize, and Monitor), and the two subscale (Inhibit, Emotional Control) and three index scores all demonstrated excellent internal consistency (BRI, MI, GEC).

Interrater-Reliability

Inter-rater reliability ICC for the BRIEF subscale and index scores are presented in Table 2. All BRIEF subscale and index scores demonstrated statistically significant ICC values. The ICC estimates ranged from poor (values less than 0.5) to moderate (values between 0.5 and 0.75) (Koo & Li, 2016). These ICC thus demonstrate poor to good agreement among parent- and teacher-ratings on the measures of executive functioning.

Gender, Age, and IQ Comparisons

Gender.—Mean scores for BRIEF parent- and teacher-report subscale and index scores for males and females are presented in Table 5. Gender differences were demonstrated on the BRIEF subscale of Parent Plan/Organize ($t[68] = -2.61$, $p = .01$), with females presenting with higher t-scores than males. However, there were no statistically significant gender differences on the percent of children reaching clinical threshold on any of the BRIEF subscales.

Gender differences were demonstrated on the BRIEF Teacher subscales of Inhibit ($t[55] = -2.39$, $p = .02$), Organization of Materials ($t[55] = -2.35$, $p = .02$), Monitor ($t[55] = -2.96$, $p = .00$) and the Metacognitive Index ($t[51] = -2.08$, $p = .04$), with females presenting with

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higher t-scores than males. There was a statistically significant gender difference on the percent of children reaching clinical threshold on the BRIEF Teacher Monitor subscale (χ^2 = 3.92, p = .05), with females being rated as being more likely to present with clinically significant problems with monitoring their work.

Age.—Correlation between BRIEF parent- and teacher-report subscale and index scores and age are presented in Table 5. Age correlated with the BRIEF Parent Monitor subscale (r = .22, p = .04). Older children with Down syndrome were reported by their parent to have more challenges with monitoring their work.

Age correlated with several BRIEF Teacher scores, including Shift (r = .31, p = .02), Initiate (r = .35, p = .01), Monitor (r = .31, p = .02), and the Metacognitive Index(r = .28, p = .04). Older children with Down syndrome were reported by their teachers to have more challenges with shifting between tasks, starting their work, and monitoring their work.

IQ.—Mean scores for BRIEF parent- and teacher-report subscale and index scores for children with IQ scores above or below the median of 42 are presented in Table 5. There were no statistically significant IQ group differences on any of the BRIEF parent-report subscales.

IQ group differences were demonstrated on the BRIEF Teacher subscale of Organization of Materials (t[54] = −2.26, p = .03), with higher functioning children demonstrating more teacher-reported challenges with organizing school materials.

With parent-report, with a few exceptions, the BRIEF normative data appears to control for gender or age variation among children with Down syndrome. With teacher-report, there is greater impact of demographics on select subscales of the BRIEF.

Discussion

The BRIEF was developed for typically developing children. Despite not being psychometrically evaluated with children with Down syndrome, both the BRIEF and BRIEF-P are commonly used in research and clinic with children with Down syndrome. We evaluated the psychometric properties of the BRIEF among school-age children with Down syndrome. The BRIEF subscale and index scores demonstrate sound psychometric properties of internal consistency for both parent- and teacher-rating forms. Normal distributions were found for subscale and index t-scores on both parent- and teacher-reports of the BRIEF. Regarding agreement among parent and teacher raters, the BRIEF subscale and index scores demonstrate poor to good inter-rater reliability, consistent with other findings of lower rates of inter-rater reliability across ratings from parents and teachers, and higher than the inter-rater reliability reported in the normative sample of the BRIEF (Achenbach, McConaughy, & Howell, 1987; Gioia, 2000). This pattern suggests that the BRIEF demonstrates moderate reliability among school-age children with Down syndrome.

With regard to characterizing our sample, we found that a substantial proportion of children were rated as having clinically significant problems with executive function across the home and school environments. These findings are consistent with results from previous studies

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that used performance and rater-based measures of executive function in this population (for review see Lee et al., 2011). Difficulties with executive functioning were reported to be more prevalent at school on teacher-reports, specifically with challenges noted from 36% (Organization of Materials) to 75% (Working Memory) on teacher-reports. In contrast, only 4–11% of typically developing children are reported to exhibit t-scores above 65 on any BRIEF subscales (Gioia, 2000). These patterns of findings illustrate two important conclusions. First, the rates of concern for executive functioning are higher in children with Down syndrome than their typically developing peers. And second, similar to findings from typically developing children, these findings may reflect different expectations across settings, such as the increased demands for planning, organization, and self-monitoring at higher grade levels. The discrepancy between typically developing children and children with Down syndrome may be particularly relevant in educationally inclusive settings. At school, children with Down syndrome may be more frequently expected to start tasks, to shift between tasks, or to mentally manipulate information on tasks. There is also less flexibility in the schedule and ability to accommodate the child as might happen in the home. Parents may be better able to address difficulties than a teacher responsible for multiple students. Sources of frustration, with peers, with tasks, with transitions may also be more common at schools than at home, resulting in more challenges with emotional control. Thus, challenges with executive functioning may be more observable to teachers or reflective of the different setting and demands. This elevated pattern of concerns with executive function at school highlights the need to focus clinical interventions on supporting the executive functioning of children with Down syndrome (Conners, Rosenquist, Arnett, Moore, & Hume, 2008; Conners, Rosenquist, & Taylor, 2001). Working memory was a noted concern for 75% of children at schools and 51% of children at home. Modifications, interventions and preventative strategies which support working memory may facilitate task compliance, task engagement and learning for school-age children with Down syndrome (Bennett, Holmes, & Buckley, 2013; Conners et al., 2008; Conners et al., 2001; Valencia-Naranjo & Robles-Bello, 2017). These types of interventions are currently being developed for youth with autism spectrum disorders (R21 HD 090334).

The most common areas of executive functioning noted to be concerning for children with Down syndrome in this sample included parent-reports of inhibitory control, shifting, working memory and task monitoring, and teacher-reports of inhibitory control, initiating on tasks, working memory and task monitoring. The high rate of concerns with inhibitory control and working memory were expected and are supported by the literature (Daunhauer et al., 2014; Lee et al., 2011). The high rate of concerns with initiating on tasks has not been noted as frequently in the literature. Clinically, this area of concern makes sense in the context of the slow processing speed of children with Down syndrome and behavioral concerns of noncompliance and motivation. Children with Down syndrome may take longer to understand tasks, and thus may present as struggling with initiation. They may also lack motivation in learning a specific topic if it is not contextually based or immediately reinforcing and therefore motivating to learn (Stein, 2016). These findings suggest a need for clearer instruction and patience in academic supports, modification of work to incorporate context and real world application, and also instruction for the child to request assistance if needed to initiate tasks more independently.

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The BRIEF provides t-scores based on age- and gender-normative data. After using the normative data conversions, some differences in t-scores were present for gender, age, and IQ. In this sample, females were reported to have more concerns with parent-reports of planning and teacher-reports of inhibitory control, organizing materials, and task-monitoring. This finding does not replicate gender differences in other preliminary examinations, where males have been reported to have more concerns on the Shift subscale (Maiman et al., 2017). Children in this sample with IQ scores above the median split demonstrated more concerns with teacher-reports of organizing materials. Again, these findings do not replicate preliminary findings where the inverse was found using parent-reports (Maiman et al., 2017). The inconsistent pattern of gender and IQ impacting t-scores on the BRIEF warrants future evaluation with a larger study population.

As the BRIEF had not been psychometrically evaluated with individuals with Down syndrome, others had adopted use of the BRIEF-P and scoring based on mental age equivalents (D'Ardhuy et al., 2015; Lee et al., 2011). The current findings suggest that for school-age children the BRIEF is an appropriate version to use. For research, studies can select whether to use the BRIEF-P with modified scoring, or the BRIEF. For clinic, it is recommended that the BRIEF is used to ensure an accurate comparison group for evaluating scores above clinical thresholds.

There are several future directions for use of the BRIEF in children with Down syndrome. As executive functioning was a concern for a substantial number of children with Down syndrome, understanding the impact of these challenges on other domains is warranted. Challenges, and patterns of challenges, with executive functioning should be further explored to better understand their impact on the adaptive skills, social functioning, language acquisition, and maladaptive behavior of children with Down syndrome (Edgin et al., 2010; Gilmore & Cuskelly, 2009; Jacola et al., 2014). Challenges with shifting between tasks combined with challenges with emotional control may contribute to more maladaptive behaviors in children with Down syndrome. Challenges with initiating tasks may contribute to noncompliant behaviors. Understanding the function of deficits in any of these domains will better inform treatment interventions.

Future research is also needed to explore different profiles of challenges in executive functioning based on the presence of comorbid medical conditions. Children with Down syndrome are at greater risk of having childhood leukemia, sleep apnea, congenital heart defects, and hypothyroidism (Bull & Genetics, 2011). Any of these comorbidities could put the child at risk for additional delays in neurodevelopment and thus identify children in need of additional intervention supports.

The current study has several limitations. Test-retest reliability was not assessed, nor was validity of the subscales or indices. Others have evaluated individual subscales and reported sound evidence of re-test reliability of these subscales and sound convergent validity for select subscales (Edgin et al., 2017; Edgin et al., 2010). However, as standardized measures of executive functioning appropriate for children with Down syndrome are not established for all subscales on the BRIEF, further evaluations of convergent validity would be premature (Esbensen et al., 2017). We did not correct for multiple comparisons. Thus,

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replication of the presented psychometric properties of the BRIEF among children with Down syndrome is necessary. Despite these limitations, there are numerous strengths to the current study. Our study is the first psychometric evaluation of the BRIEF in school-age children with Down syndrome, including both clinic and community samples. Inclusion of community and clinic samples helps ensure a more representative sample of children with Down syndrome and variability in presenting concerns.

This study contributes to our understanding of measurement of executive functioning through informant-based rating scales in children with Down syndrome. The findings of this study have important implications for accurately assessing the subcomponents of executive functioning and expanding measurement options for evaluating outcomes in children with Down syndrome in future clinical trials and treatment interventions (Esbensen et al., 2017; NICHD, 2015). Continuing to understand how the BRIEF may be used to assess for changes over time or detecting treatment related change is needed. In addition, understanding how the BRIEF may be used as a screener to inform environmental accommodations at school and support learning of individual children with Down syndrome is also warranted. Altogether, our findings suggest that the BRIEF is appropriate and meaningful for use with school-age children with Down syndrome.

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References

Achenbach TM, McConaughy SH, & Howell CT (1987). Child/adolescent behavioral and emotional problems: Implications of cross-informant correlations for situational specificity. Psychological bulletin, 101(2), 213–232. [PubMed: 3562706]

Baddeley A (1998). Recent developments in working memory. Current opinion in neurobiology, 8(2), 234–238. [PubMed: 9635207]

Baddeley A, & Jarrold C (2007). Working memory and Down syndrome. Journal of Intellectual Disability Research, 51(12), 925–931. [PubMed: 17990999]

Bennett SJ, Holmes J, & Buckley S (2013). Computerized memory training leads to sustained improvement in visuospatial short-term memory skills in children with Down syndrome. American journal on intellectual and developmental disabilities, 118(3), 179–192. [PubMed: 23734613]

Borella E, Carretti B, & Lanfranchi S (2013). Inhibitory mechanisms in Down syndrome: Is there a specific or general deficit? Research in developmental disabilities, 34(1), 65–71. [PubMed: 22940160]

Bull MJ, & Genetics C. o. (2011). Health supervision for children with Down syndrome. Pediatrics, 128(2), 393–406. doi:10.1542/peds.2011-1605 [PubMed: 21788214]

Campbell C, Landry O, Russo N, Flores H, Jacques S, & Burack JA (2013). Cognitive flexibility among individuals with Down syndrome: assessing the influence of verbal and nonverbal abilities. American journal on intellectual and developmental disabilities, 118(3), 193–200. [PubMed: 23734614]

Carney DP, Brown JH, & Henry LA (2013). Executive function in Williams and Down syndromes. Research in developmental disabilities, 34(1), 46–55. [PubMed: 22940158]

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Carney DP, Henry LA, Messer DJ, Danielsson H, Brown JH, & Rönnberg J (2013). Using developmental trajectories to examine verbal and visuospatial short-term memory development in children and adolescents with Williams and Down syndromes. Research in developmental disabilities, 34(10), 3421–3432. [PubMed: 23920025]

Conners F, Rosenquist C, Arnett L, Moore M, & Hume L (2008). Improving memory span in children with Down syndrome. Journal of Intellectual Disability Research, 52(3), 244–255. [PubMed: 18261023]

Conners F, Rosenquist C, & Taylor L (2001). Memory training for children with Down syndrome. Down Syndrome Research and Practice, 7(1), 25–33.

Costanzo F, Varuzza C, Menghini D, Addona F, Gianesini T, & Vicari S (2013). Executive functions in intellectual disabilities: a comparison between Williams syndrome and Down syndrome. Research in developmental disabilities, 34(5), 1770–1780. [PubMed: 23501586]

D'Ardhuy XL, Edgin JO, Bouis C, de Sola S, Goeldner C, Kishnani P, … Khwaja O (2015). Assessment of cognitive scales to examine memory, executive function and language in individuals with Down syndrome: implications of a 6-month observational study. Frontiers in Behavioral Neuroscience, 9, 300. [PubMed: 26635554]

Daunhauer LA, Fidler DJ, Hahn L, Will E, Lee NR, & Hepburn S (2014). Profiles of everyday executive functioning in young children with Down syndrome. American journal on intellectual and developmental disabilities, 119(4), 303–318. [PubMed: 25007296]

Dykens E, Hodapp R, & Finucane BM (2000). Genetics and mental retardation syndromes: A new look at behavior and interventions: Paul H Brookes Publishing.

Edgin JO, Anand P, Rosser T, Pierpont EI, Figuero C, Hamilton D, … Sherman S (2017). The Arizona Cognitive Test Battery for Down Syndrome: Test-retest reliability and practice effects. American Journal of Intellectual and Developmental Disabilities, 122(3), 215–234.

Edgin JO, Mason GM, Allman MJ, Capone GT, DeLeon I, Maslen C, … Nadel L (2010). Development and validation of the Arizona Cognitive Test Battery for Down syndrome. Journal of neurodevelopmental disorders, 2, 149–164. [PubMed: 21274406]

Esbensen AJ, & Hoffman E (2017). Sleep Problems in Children with Down Syndrome: The Impact of Raters on Outcomes. Paper presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Antonio, TX.

Esbensen AJ, Hooper SR, Fidler D, Hartley SL, Edgin J, d'Ardhuy XL, … Urv T (2017). Outcome measures for clinical trials in Down syndrome. American journal on intellectual and developmental disabilities, 122(3), 247–281. [PubMed: 28452584]

Fry AF, & Hale S (1996). Processing speed, working memory, and fluid intelligence: Evidence for a developmental cascade. Psychological science, 7(4), 237–241.

Gilmore L, & Cuskelly M (2009). A longitudinal study of motivation and competence in children with Down syndrome: Early childhood to early adolescence. Journal of Intellectual Disability Research, 53(5), 484–492. [PubMed: 19302472]

Gioia GA (2000). Behavior Rating Inventory of Executive Function: Professional Manual. Odessa, FL: Psychological Assessment Resources, Incorporated.

Gioia GA, Espy KA, & Isquith PK (2003). BRIEF-P: Behavior Rating Inventory of Executive Function--Preschool Version: Professional Manual. Odessa, FL: Psychological Assessment Resources.

Jacola LM, Hickey F, Howe SR, Esbensen A, & Shear PK (2014). Behavior and adaptive functioning in adolescents with Down syndrome: Specifying targets for intervention. Journal of Mental Health Research in Intellectual Disabilities, 7(4), 287–305. [PubMed: 28539987]

Kishnani PS, Heller JH, Spiridigliozzi GA, Lott I, Escobar L, Richardson S, … McRae T (2010). Donepezil for treatment of cognitive dysfunction in children with Down syndrome aged 10–17. American Journal of Medical Genetics Part A, 152(12), 3028–3035.

Koo TK, & Li MY (2016). A guideline of selecting and reporting intraclass correlation coefficients for reliability research. Journal of chiropractic medicine, 15(2), 155–163. [PubMed: 27330520]

Landry O, Russo N, Dawkins T, Zelazo PD, & Burack JA (2012). The impact of verbal and nonverbal development on executive function in Down syndrome and Williams syndrome. Journal on Developmental Disabilities, 18(2).

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Lanfranchi S, Jerman O, Dal Pont E, Alberti A, & Vianello R (2010). Executive function in adolescents with Down Syndrome. Journal of Intellectual Disability Research, 54, 308–319. [PubMed: 20202074]

Lee NR, Fidler DJ, Blakeley-Smith A, Daunhauer L, Robinson C, & Hepburn SL (2011). Caregiver report of executive functioning in a population-based sample of young children with Down syndrome. American journal on intellectual and developmental disabilities, 116(4), 290–304. [PubMed: 21740257]

Maiman M, Hamn e., T., Udhnani M, Anand P, Olasen LS, Giedd JN, … Lee NR (2017). Differential effects of sex and intellectual ability level on “cool” and “hot” executive function impairments in youth with Down syndrome. Paper presented at the Gatlinburg Conference on Research and Theory in Intellectual and Developmental Disabilities, San Antonio, TX.

NICHD. (2015). Outcome measures for clinical trials in individuals with Down syndrome. Retrieved from https://www.nichd.nih.gov/about/meetings/2015/Documents/DS_outcomes_meeting_summary.pdf

Pennington BF, Moon J, Edgin J, Stedron J, & Nadel L (2003). The neuropsychology of Down syndrome: evidence for hippocampal dysfunction. Child development, 74(1), 75–93. [PubMed: 12625437]

Pritchard A, Kalback S, McCurdy M, & Capone GT (2015). Executive functions among youth with Down Syndrome and co-existing neurobehavioural disorders. Journal of Intellectual Disability Research, 59(12), 1130–1141. [PubMed: 26369785]

Rowe J, Lavender A, & Turk V (2006). Cognitive executive function in Down’s syndrome. British Journal of Clinical Psychology, 45(1), 5–17. [PubMed: 16480563]

Silverman W (2007). Down syndrome: Cognitive phenotype. Mental retardation and developmental disabilities research reviews, 13(3), 228–236. [PubMed: 17910084]

Stein D (2016). Supporting Positive Behavior in Children and Teens with Down Syndrome. Bethesda, MD: Woodbine House.

Valencia-Naranjo N, & Robles-Bello MA (2017). Learning potential and cognitive abilities in preschool boys with fragile X and Down syndrome. Research in developmental disabilities, 60, 153–161. [PubMed: 27984817]

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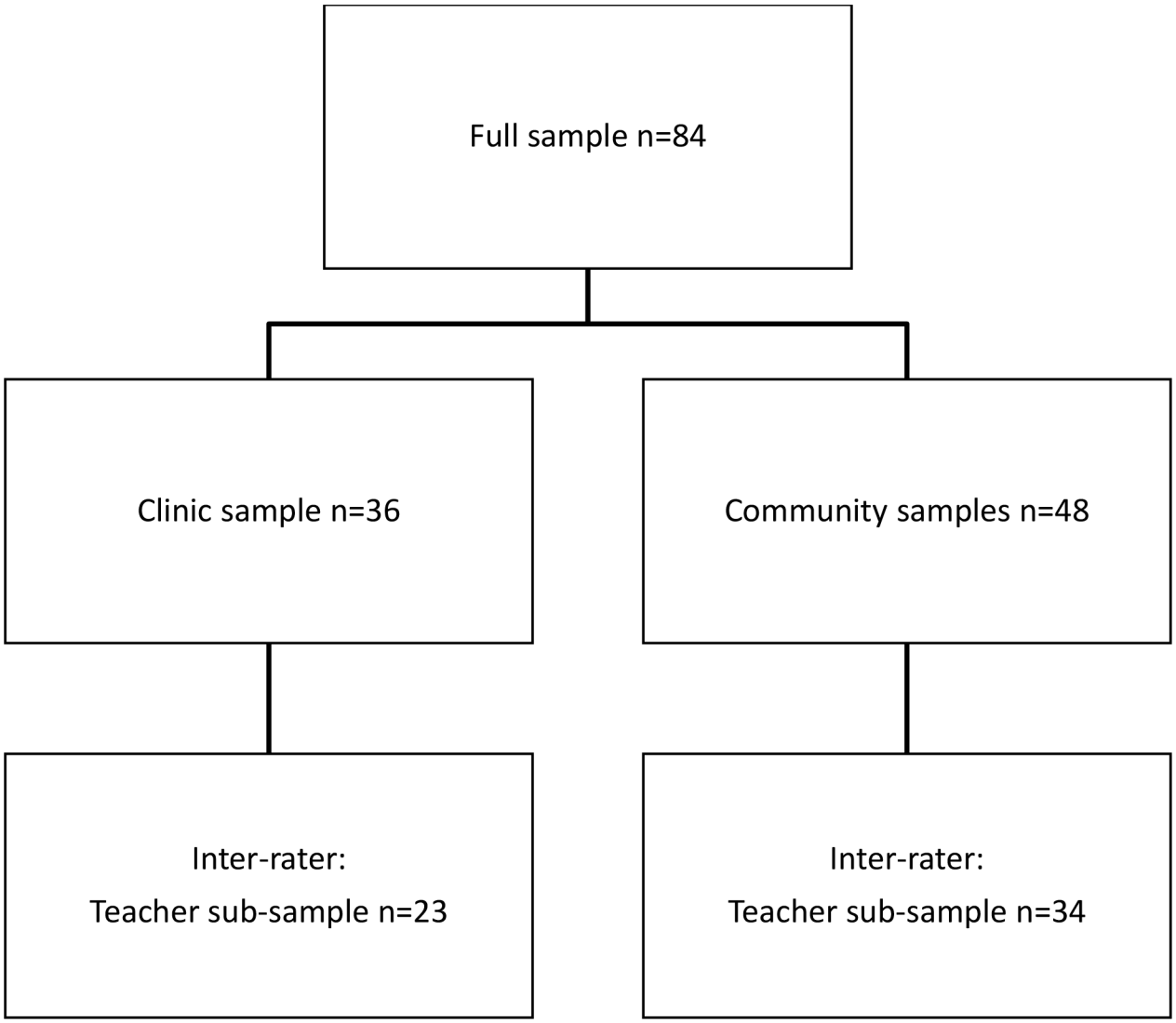

Figure 1.
Flow diagram of samples included in analysis

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Table 1.

Mean standard scores on measures of cognition.

	n	Mean (SD)	Range
DAS-II General Conceptual Ability	23	43.83 (10.36)	25–65
Wechsler Intelligence Scale for Children-IV	5	47.80 (8.20)	40–60
Stanford-Binet-5	8	42.88 (2.70)	40–47
Kaufman Brief Test of Intelligence	48	43.50 (4.89)	40–57

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Table 2.

Psychometric properties of BRIEF parent and teacher subscales.

	N	Mean (SD)	Range	Clinical concern	Alpha	Inter-rater reliability
BRIEF Parent Report						
Inhibit	84	63.2 (12.2)	38–97	44.0%	.88	.57**
Shift	84	62.0 (13.5)	36–92	38.1%	.78	.46*
Emotional Control	84	53.7 (12.0)	35–88	15.5%	.88	.58**
BRI	82	60.6 (12.6)	34–97	35.4%	.92	.55**
Initiate	82	59.0 (10.1)	35–86	30.5%	.67	.38*
Working Memory	84	64.6 (9.5)	36–84	51.2%	.83	.45*
Plan/Organize	70	60.1 (11.9)	37–95	35.7%	.84	.67**
Organization of Materials	82	52.9 (9.2)	33–73	9.8%	.84	.37*
Monitor	82	64.7 (11.2)	32–91	56.1%	.82	.64**
MI	68	62.1 (10.3)	34–86	45.6%	.94	.57**
GEC	70	62.4 (10.7)	35–81	41.4%	.96	.62**
BRIEF Teacher Report						
Inhibit	57	73.1 (16.4)	44–120	64.9%	.94	-
Shift	57	66.5 (16.7)	42–114	52.6%	.89	-
Emotional Control	57	66.3 (16.5)	43–109	50.9%	.92	-
BRI	57	71.0 (16.5)	43–106	61.4%	.96	-
Initiate	57	69.8 (11.4)	41–92	70.2%	.81	-
Working Memory	57	74.1 (13.3)	50–106	75.4%	.89	-
Plan/Organize	53	63.9 (12.6)	40–95	49.1%	.85	-
Organization of Materials	57	63.7 (16.6)	42–116	36.8%	.76	-
Monitor	53	72.5 (14.4)	41–102	68.4%	.87	-
MI	53	71.3 (12.8)	44–100	69.8%	.96	-
GEC	53	72.4 (14.2)	45–103	71.7%	.97	-

*p < .05

**p < .01

BRI = Behavioral Regulation Index, MI – Metacognition Index, GEC – Global Executive Composite

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Table 3.

Inter-correlation of BRIEF parent-report subscales (n=84).

		1	2	3	4	5	6	7	8	9	10
1	Inhibit	-									
2	Shift	.49**	-								
3	Emotional Control	.71**	.61**	-							
4	BRI	.87**	.80**	.91**	-						
5	Initiate	.39**	.46**	.46**	.50**	-					
6	Working Memory	.49**	.41**	.34**	.47**	.62**	-				
7	Plan/Organize	.41**	.28*	.35**	.40**	.59**	.48**	-			
8	Organization of Materials	.46**	.49**	.51**	.56**	.49**	.46**	.44**	-		
9	Monitor	.53**	.36**	.44**	.51**	.68**	.64**	.71**	.42**	-	
10	MI	.58**	.54**	.52**	.62**	.83**	.81**	.84**	.68**	.88**	-
11	GEC	.80**	.70**	.77**	.87**	.77**	.78**	.72**	.72**	.81**	.92**

* p < .05

** p < .01

BRI = Behavioral Regulation Index, MI – Metacognition Index, GEC – Global Executive Composite

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Table 4.

Inter-correlation of BRIEF teacher-report subscales (n=57).

		1	2	3	4	5	6	7	8	9	10
1	Inhibit	-									
2	Shift	.58**	-								
3	Emotional Control	.79**	.72**	-							
4	BRI	.90**	.83**	.94**	-						
5	Initiate	.58**	.52**	.47**	.59**	-					
6	Working Memory	.63**	.54**	.42**	.60**	.78**	-				
7	Plan/Organize	.63**	.62**	.59**	.68**	.68**	.69**	-			
8	Organization of Materials	.44**	.29*	.38**	.42**	.54**	.58**	.64**	-		
9	Monitor	.83**	.56**	.68**	.78**	.68**	.68**	.68**	.49**	-	
10	MI	.75**	.64**	.59**	.73**	.85**	.89**	.88**	.75**	.86**	-
11	GEC	.86**	.80**	.78**	.90**	.81**	.83**	.85**	.64**	.88**	.95**

* p < .05

** p < .01

BRI = Behavioral Regulation Index, MI – Metacognition Index, GEC – Global Executive Composite

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Table 5.

Gender and IQ difference on BRIEF parent and teacher subscales and correlation of subscales with age.

		Males		Females		Correlation with age	IQ ≤ 42	IQ ≥ 43
	n	Mean (SD)	Clinical concern	Mean (SD)	Clinical concern		Mean (SD)	Mean (SD)
BRIEF Parent Report								
Inhibit	84	63.6 (13.3)	45.1%	62.6 (10.3)	42.4%	.20	62.2 (12.8)	64.2 (11.8)
Shift	84	63.1 (14.9)	43.1%	60.2 (11.0)	30.3%	.16	61.0 (12.8)	63.3 (14.5)
Emotional Control	84	53.7 (13.0)	17.6%	53.6 (10.6)	12.1%	.15	52.1 (11.0)	55.4 (13.4)
BRI	82	61.3 (13.9)	40.8%	59.6 (10.5)	27.3%	.19	59.3 (11.5)	62.0 (13.8)
Initiate	82	57.4 (10.8)	28.6%	61.3 (8.7)	33.3%	.18	58.9 (9.4)	58.8 (11.1)
Working Memory	84	64.1 (10.1)	52.9%	65.4 (8.4)	48.5%	.12	64.4 (9.5)	64.3 (9.3)
Plan/Organize	70	57.2 (11.1)[a]	28.6%	64.5 (12.0)[a]	46.4%	.09	57.4 (11.1)	61.6 (11.2)
Organization of Materials	82	53.4 (9.7)	10.2%	52.1 (8.4)	9.1%	−.13	51.8 (9.5)	53.8 (8.8)
Monitor	82	62.9 (11.2)	57.1%	67.3 (10.9)	54.5%	.22*	64.3 (10.5)	64.6 (11.7)
MI	68	60.6 (10.9)	47.5%	64.2 (9.3)	42.9%	.16	60.6 (9.3)	62.8 (10.6)
GEC	70	61.6 (11.5)	42.9%	63.5 (9.5)	39.3%	.23	60.8 (9.3)	63.3 (11.9)
BRIEF Teacher Report								
Inhibit	57	69.4 (11.9)[a]	59.5%	79.8 (21.1)[a]	75.0%	.10	70.8 (15.6)	74.3 (16.9)
Shift	57	66.3 (17.5)	48.6%	66.8 (15.6)	60.0%	.31*	68.8 (19.5)	64.0 (13.8)
Emotional Control	57	65.2 (15.5)	48.6%	68.3 (18.6)	55.0%	.14	63.8 (17.4)	68.1 (15.7)
BRI	57	69.0 (15.1)	59.5%	74.7 (18.6)	65.0%	.18	69.7 (16.9)	71.4 (16.1)
Initiate	57	68.5 (9.7)	64.9%	72.2 (14.0)	80.0%	.35**	69.3 (11.2)	70.1 (12.0)
Working Memory	57	71.9 (11.4)	73.0%	78.2 (15.8)	80.0%	.22	74.4 (12.2)	73.7 (14.7)
Plan/Organize	53	62.6 (11.1)	47.2%	66.6 (15.3)	52.9%	.17	60.8 (12.5)	66.4 (12.3)
Organization of Materials	57	60.1 (14.2)[a]	29.7%	70.5 (19.0)[a]	50.0%	.18	58.7 (11.3)[a]	68.5 (19.7)[a]
Monitor	57	68.6 (10.9)[a]	59.5%[b]	79.6 (17.3)[a]	85.0%[b]	.31*	70.3 (13.9)	73.9 (14.8)
MI	53	68.9 (10.3)[a]	66.7%	76.5 (16.1)[a]	76.5%	.28*	69.2 (10.9)	73.0 (14.5)
GEC	53	70.4 (12.6)	69.4%	76.5 (16.8)	76.5%	.26	70.4 (13.2)	73.7 (15.2)

*p < .05

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** $p < .01$

[a] significant group difference

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