**Symposium Title:** Measurement in Down Syndrome: Examining Heterogeneity in Developmental Syndromes

**Chair:** Aaron Dallman¹

**Discussant:** Anna Esbensen²

**Overview:** Down Syndrome (DS), with an estimated prevalence of 6.7 per 10,000 inhabitants, is a neurodevelopmental disorder caused by altered protein expression of genes in chromosome 21 (De Graaf, Buckley, & Skotko, 2017). While a number of studies have investigated comorbid health conditions within this group, there is still a need to characterize the behavioral phenotype and experiences of young children and adolescents with DS. Further, current outcome measures are problematic in this population due to significant floor effects, inability to detect change over time, and impracticality leading to decreased understanding of behavioral phenotypes and heterogeneity in the DS population. Validation of current measures and development of new ones that address the aforementioned concerns will serve to better identify subgroups within the DS population and evaluate the effects of pharmacologic and behavioral interventions. This graduate student symposium features three presentations that will address a few salient aspects of measurement of the Down syndrome phenotype in children. The first presentation will use a novel measure, the Behavioral Inflexibility Scale, to investigate rigid and inflexible behavior within the DS population. The second presentation will examine the executive functioning profile of children with DS by identifying specific EF deficits within this population and EF changes over time. Finally, the third presentation seeks to identify reliable peer victimization, emotion regulation, and self-esteem self-report measures.

**Paper 1 of 3**

**Paper Title:** Behavioral Inflexibility: Comparison of Down Syndrome and Autism

**Authors:** Aaron Dallman¹, Clare Harrop³, Brian Boyd³, Luc Lecavalier⁴, James Bodfish⁵

**Introduction:** Down Syndrome (DS) is a genetic syndrome associated with distinct health characteristics such as congenital heart disease and duodenal stenosis. In addition, emerging research suggests that DS is characterized by a unique behavioral phenotype that distinguishes the DS population from other intellectual/developmental disorders. Recent studies have shown that individuals with DS demonstrate greater inflexibility than the typically developing population, but less behavioral inflexibility than peers with autism spectrum disorders (Green et al., 2006). This study seeks to apply a novel behavioral inflexibility instrument, Behavioral Inflexibility Scale (BIS), to the DS population, in order to further characterize the behavioral inflexibility of this population.

**Methods:** Participants included 75 caregivers of children with DS (35 males and 40 females; 3 to 17 years), 142 caregivers of children with autism spectrum disorder (ASD; 117 males and 25 females; 3 to 17 years), and 92 (61 males and 31 females; 3 to 15 years) caregivers of typically developing (TD) children. Of the DS participants, 49 (65%) had a diagnosis of comorbid mental retardation/intellectual disability. Caregivers completed the Behavioral Inflexibility Scales (BIS; Boyd et al., 2018). To account for age differences between groups, age was controlled for in the analyses. To test the sensitivity of the BIS, mean scores from DS, ASD, and TD populations were compared using linear regression. T-tests were conducted, controlling for multiple comparisons using a Bonferroni correction, to test for differences between DS and ASD groups amongst individual items.

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Results: The BIS demonstrated strong internal consistency (alpha=0.95) in this sample. DS participants (mean=1.22, sd=.90) demonstrated less inflexibility (beta=0.95, p<.01) than the ASD group (mean=2.16, sd=1.07), but the DS group demonstrated more inflexibility (beta=-0.98, p>.01), than the TD participants (mean=.23, sd=.35). However, inflexibility did not differ by age (beta=-0.03, p=.13). There was no interaction effect with gender and BIS score (p>.05), but there was a significant interaction effect (beta=0.07, p=.02) with age and BIS score suggesting that, at older ages, the difference between ASD and DS score in the BIS is reduced. No significant differences were found between DS and ASD groups for the following items and thus these items represent a common subset of inflexibility symptoms amongst individuals with ASD and DS: resists changes to the way he usually does things, takes a long time to get comfortable in new situations, has trouble coming up with new ways of doing things, and, hard to redirect from things he is doing. All other items showed statistically significant differences (p<.001) and were endorsed more in the ASD sample.

Discussion: The results suggest that the BIS could be a promising tool for identifying individuals with DS who show behavioral inflexibility. The scale appears to have discriminant validity as analysis demonstrated differences between DS groups and individuals with ASD as well as TD. These results echo previous findings suggesting that individuals with DS demonstrate decreased behavioral inflexibility compared to ASD peers, but increased inflexibility relative to neurotypical peers. Further, the lack of observed differences between age groups within the DS population suggests that behavioral inflexibility may remain a core component of the behavioral phenotype of DS across childhood.

References/Citations:

Paper Title: Measuring Executive Functioning Profiles and Development in Young Children with Down Syndrome

Authors: Amanda Dimachkie\textsuperscript{6}, Kyle Sterrett\textsuperscript{6}, Lauren Baczewski\textsuperscript{6}, Alison Holbrook\textsuperscript{6}, Connie Kasari\textsuperscript{6}

Introduction: Children with Down Syndrome (DS) are consistently impaired in executive functioning (EF) skills beyond what would be expected when accounting for mental age (MA; Daunhauer et al., 2014) particularly across domains such as working memory and planning/organization (Loveall et al., 2017). Further, deficits in language abilities appear to be related to executive functioning impairments (Grieco et al., 2015). However, few studies have studied the development of EF skills over time in very young children with DS as measured by parent report.

Methods: 39 children with DS (mean CA =41.6 months; mean MA = 23.94 months) were recruited to participate in an early social communication intervention (JASPER/EMT). At entry, three-, and six-months timepoints parents completed the BRIEF-P. T-scores were calculated for each composite domain: Emerging Metacognition (EMI), Inhibitory Self Control Index (ISCI) and Flexibility Index (FI). Children were also administered the Preschool Language Scale-5 (PLS-5) and Mullen Scales of Early Learning at each timepoint. Three separate linear mixed models were used to evaluate the relationship between time and children’s scores on EMI, ISCI & FI domains. Fixed effects included PLS-5 Auditory Comprehension (AC) scores and timepoint (Entry, 3-month, 6-month) and a random effect was included for children’s intercept. For baseline data, multiple regression was used to evaluate the relationship between EMI, ISCI and FI scores at entry, PLS-5 AC and Expressive Communication (EC) scores, mental age and chronological age.

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Results: Means and standard deviations for BRIEF-P subdomains are presented in table 1. EMI but not Fl or ISCI scores, were clinically elevated across all three timepoints. Across the three subdomains there was no effect of time on BRIEF-P scores (Wald chi-squares p > .05), indicating stability over time. For the baseline models, a significant regression equation was found for ISCI (F(4,30) = 4.38, p < .01) with PLS AC and CA emerging as predictors of ISCI scores at baseline (p-values < .05). A significant regression equation was also found for Fl (F(4,30) = 7.71, p < .01) with only CA emerging as a significant predictor (p < .01). The regression equation did not reach significance for EMI F(4,30) = 1.99, p < .12 although a similar pattern to the ISCI emerged with PLS AC emerging as a significant predictor.

Discussion: Results provide further evidence for a working memory deficit in DS, extending this finding to young children. Consistent with extant literature regarding older individuals with DS (Grieco et al., 2015), EF abilities appear to be stable over time in toddlers. The sample showed marked deficits in both working memory and planning/organization, which are two skill domains essential for later academic success (Alloway & Alloway, 2010). Auditory comprehension and chronological age emerged as unique predictors of children’s EF abilities. The stability of the executive functioning impairments over time suggests targeted interventions are needed to improve EF skills in individuals with DS.

References/Citations:

Table 1.

<table>
<thead>
<tr>
<th>BRIEF-P Subdomain Scores Across Time</th>
<th>Entry</th>
<th>Exit</th>
<th>FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emergent Metacognition</td>
<td>70.06 (12.82)</td>
<td>69.30 (11.67)</td>
<td>68.97 (12.74)</td>
</tr>
<tr>
<td>Flexibility</td>
<td>58.69 (15.94)</td>
<td>57.24 (12.91)</td>
<td>55.26 (13.82)</td>
</tr>
<tr>
<td>Inhibitory Self-Control</td>
<td>60.22 (12.86)</td>
<td>60.76 (13.78)</td>
<td>58.42 (12.38)</td>
</tr>
</tbody>
</table>

Paper 3 of 3

Paper Title: The Measurement of Peer Victimization, Emotion Regulation, and Self-Esteem in Adolescents with Down Syndrome: A Pilot Study

Authors: Jenna Reardanz\(^7\), Frances A. Conners\(^7\), Kristina L. McDonald\(^7\)

Introduction: Victimization, or being perpetually harassed, ridiculed, and the target of aggression, is something that is a particular issue for typically developing (TD) individuals during adolescents and is thought to be influenced by one’s self-esteem (SE) and emotion regulation (ER) ability (Troop-Gordon & Unhjem, 2018; Godleski, et al., 2015; Glenn, 2001). However, very little is known about victimization in adolescents with Down syndrome (DS). Indeed, very few self-report victimization measures have been shown to be reliable in this population. In those with DS, ER and SE are typically measured through other-report and/or

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tasks; again, there are few reliable self-report measures for these constructs in this population. Therefore, this study focused on finding reliable self-report measures for each of these constructs.

**Methods:** 23 adolescents with DS between the ages of 11 and 18 years ($M_{age} = 14.13; SD = 2.46; PPVT-4 M_{test\_age} = 5.29; SD = 1.49; 91.3\%$ White Non-Hispanic; $56.6\%$ female) completed measures addressing SE, victimization, and ER. Participants completed modified versions of the ER Index for Children and Adolescents (ERICA; MacDermott, et al., 2010), the Children’s Social Experience Questionnaire (CSEQ; Crick & Grot彼得, 1996), and an adapted Rosenberg Self-Esteem Measure (Dagnan & Sandhu, 1999). We simplified language (word choice and grammar) and modified the response format of each of these Likert style measures, following Haynes, Gilmore, Shochet, Campbell, & Roberts (2013). The modified response format included breaking each question down into a two part response (Yes/No followed by A lot/A little), resulting in a 4-point scale. This placed less burden on memory and comprehension processes.

**Results:** Internal reliability analyses demonstrated that the victimization measure, CSEQ, was reliable for self-report in this sample ($M = 32.35; SD = 7.32, \alpha = .774$) and was approximately normally distributed. The self-report SE measure, Rosenberg, was neither internally reliable ($M = 21.65; SD = 1.69, \alpha = .467$) nor normally distributed. The SE measure was negatively skewed, indicating reports of high SE. Finally, the self-report measure of ER, the ERICA, was normally distributed, but not internally reliable ($M = 31.26; SD = 2.78, \alpha = .419$).

**Discussion:** The modified CSEQ measure seems to be a potentially reliable self-report measure to utilize in research with those with DS. However, the ERICA and the Rosenberg Self-Esteem Measure do not seem yet to be reliable, at least with the modification used in this study. Possibly, adolescents with DS are able to more reliably report their experiences (as in the CESQ) than their inner states (as in the ERICA and Rosenberg). Demonstration tasks may be more effective at tapping ER and self-esteem compared with self-report for this population. It is important to note, however, that with this small sample size (N = 23), psychometric properties cannot be fully assessed. As our sample expands, firmer conclusions will be possible. However, at this point, the modified CESQ seems a promising measure of peer victimization for adolescents with DS.

**References/Citations:**
- Spectrum Disorders, 7, 931-937.