proportion endorsing clinically significant concerns with behavior regulation (68%) relative to cognitive regulation (48%). Majority of participants was rated in the low or very low range for ABAS-3 General Adaptive Composite (72%), with a greater proportion showing problems in Practical (64%) and Conceptual Domains (64%) relative to Social Domain (44%). Among those who completed the SDQ, caregiver ratings implicate elevated Total Problems (90%) with greater concerns observed in Emotional Problems (62%), Hyperactivity (81%) and Peer Relationship difficulties (95%). After controlling for age, executive functioning difficulties were associated with weaker skills in the Conceptual (r=-0.56, p=0.003), Social (r=-0.44, p=0.028) and Practical domains (r=-0.51, p=0.009); as well as more Hyperactivity (r=0.49, p=0.025) and Conduct problems (r=0.58, p=0.007). Specifically, day-to-day challenges with executive functions were related to weaker adaptive skills in Self-Care (r=-0.54, p=0.006), Self-Direction (r=-0.53, p=0.007), and Communication (r=-0.49, p=0.01).
Conclusions: Individuals with WSS are at risk for executive functioning deficits, which in turn may impact the development of and/or day-to-day application of adaptive skills and behavior regulation. Future clinical research should further explore the development and neurophysiology of executive functions among those with WSS with multidisciplinary methods, including behavioral, cognitive and neurobiological metrics. Those working with individuals with WSS may consider executive functioning interventions, which may yield indirect benefits to self-regulation and daily use of life skills.

Categories: Genetics/Genetic Disorders
Keyword 1: adaptive functioning
Keyword 2: executive functions
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58 Parent Ratings of Internalizing and Externalizing Behaviors in Children with NF1 Across Childhood: A Longitudinal Investigation

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Objective: The purpose of the present study is to characterize the trajectory of internalizing and externalizing behaviors in children with NF1 longitudinally from the early childhood period to the school age period on a broad psychosocial norm-referenced measure using linear mixed model growth curves.
Participants and Methods: Children with NF1 (n=28) were seen at least once between the ages of 3-8 years old and then again between the ages of 9-13 years old. Parents completed the Behavior Assessment System for Children (BASC) Second Edition; the version of the BASC administered depended on age (i.e., preschool form or child form). Linear mixed model growth curve analyses were used to examine the developmental trajectories of children with NF1 on the following scales, which were selected due to findings in the literature: Externalizing Problems, Internalizing Problems, Hyperactivity, Anxiety, Depression, Attention, and Executive Function. T-scores (M=50, SD=10) were used. Higher scores indicate more challenges.
Results: By using loess lines to qualitatively describe the patterns of ratings across time, it is evident that most scales (Externalizing Problems, Internalizing Problems, Hyperactivity, Attention Problems, Executive Function) demonstrated curvilinear trajectory patterns, with scores peaking in the 8–10-year-old range, then decreasing again. However, there was no statistically significant effect of age on any of the scales. Notably, trajectories largely included standard scores within the normative range (T-scores between 45-55).
Conclusions: Overall, the models also suggest that most children with NF1 are within the average range of functioning on all scales examined across the childhood period. Furthermore, with the exception of the Depression and Anxiety scales, ratings tend to peak around the 8–10-year period, and then decrease into early adolescence. Thus, when working with patients with NF1, it may be the case that clinicians note relative increases in challenges across these domains in late childhood, though these challenges may decrease over time during this age range. Linear growth curve modeling identified that the developmental trajectories of internalizing and externalizing behaviors of children with NF1, as

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rated by parents, remain stable across the childhood period. Importantly, low power may have contributed to the lack of observed age effects. Longitudinal research would be beneficial to capture patterns that may emerge in adolescence or adulthood.

Categories: Genetics/Genetic Disorders
Keyword 1: neurofibromatosis
Keyword 2: genetic disorders
Keyword 3: everyday functioning
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59 Preliminary Findings of Semantic Interference in Learning and Memory Processes in Manifest Huntington's Disease

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Objective: To explore the usefulness of the Lowenstein-Acevedo Scales for Semantic Interference and Learning (LASSI-L) [Crocco et al, 2013], a novel memory-based cognitive stress test capitalizing on semantic interference, in Huntington's Disease (HD).
Participants and Methods: 12 healthy adults (HA) and 14 individuals with manifest HD were administered the LASSI-L as part of an annual research visit with the UCSD Huntington's Disease Clinical Research Center (HDCRC.) Participants in each group were well matched with regard to age and education. Individuals with manifest HD had an average MoCA score of 26, total functional capacity score of 10, and total motor score of 21 suggesting that they were in the early stages of HD. The LASSI-L examines different types of semantic interference that occur in the learning/encoding process. There are free and cued recall trials for two lists of semantically related words with certain trials specific to different aspects of semantic interference including proactive, retroactive, and failure to recover from proactive interference. T-tests for all recall trials and number of intrusions for each trial were conducted between HA and those with HD to examine whether HD renders one more prone to semantic interference in both encoding and retrieval memory processes.
Results: Individuals with HD recalled fewer words on average than HA across all recall trials except for the initial free recall of the first word list. HD individuals recalled significantly fewer (~1.5) words during the initial (t=-2.8, p=.005, Cohen's d=2.7) and secondary (t=-2.9, p=.003, Cohen's d=2.6) cued recall trials from the words on the first list. Individuals with HD also recalled significantly fewer words on initial free recall (t=-2.9, p=.003, Cohen's d=2.6) and cued recall trials of the second list, with the initial cued recall (t=-2.8, p=.005, Cohen's d=3.1) sensitive to proactive semantic interference and the second cued recall (t=-3.3, p=.001, Cohen's d=2.6) sensitive to failure to recover from proactive semantic interference. In addition, individuals with HD also recalled significantly fewer (~2.2) words on delayed cued recall of the first list, a measure of retroactive semantic interference, than HA (t=-4.8, p<.001, Cohen's d=2.4). Lastly, individuals with HD recalled fewer (~4.1) words than HA on delayed free recall of both word lists (t=-3.5, p<.001, Cohen's d=5.9). The groups did not differ significantly with regard to number of total intrusions per trial.
Conclusions: Overall, our study supports the usefulness of the LASSI-L for neuropsychological assessment of HD in clinical and research settings. In comparison to a demographically similar group of HA, individuals with manifest HD showed significant differences in frontally mediated retrieval processes as well as semantic interference processes that affect efficient encoding of novel information.

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Keyword 1: dementia - subcortical
Keyword 2: neuropsychological assessment
Keyword 3: movement disorders
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