Measures included were the Working Memory Index score from the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000) and the Digit Span subtest from the Wechsler Intelligence Scale for Children, Fourth Edition (WISC-IV; Wechsler, 2003) and the Wechsler Adult Intelligence Scale, Fourth Edition (WAIS-IV; Wechsler, 2008).

Results: Mean scores on parent-reported WM scores and cognitive measures of attention/WM fell within normal limits, including the Digit Span Total score (M = 48.42, SD = 6.33), Digit Span Forward score (M = 47.28, SD = 9.9.83), and Digit Span Backward score (M = 48.94, SD = 6.31). However, further analyses suggested that between 11-32% of patients had scores falling at least one standard deviation below the mean on these measures, with more than half of the sample (52.6%) identified with at least one measured weakness in attention and WM. The most commonly identified weakness (33.3% of patients) was Digit Span Forward.

Correlations between parent-reported WM issues and cognitive measures of attention and WM were generally strong, with parent report of WM significantly correlated with the Digit Span Total score (r(18) = -0.52, p=.02) and the Digit Span Forward score (r(18) = -0.51, p=.03). No correlations were found between Digit Span Backward and other measures of attention and WM.

There were no significant differences in WM scores between patients with ALL and AML. Additional analyses will examine potential contribution of medical factors (e.g., pre-HSCT treatment) to pre-HSCT performance on measures of attention and WM.

Conclusions: These results suggest that, prior to undergoing HSCT, pediatric patients present with attention and WM issues. This finding has implications for research related to neurocognitive outcomes in HSCT, indicating the need to obtain pre-HSCT cognitive data in this area in order to fully understand potential change after HSCT. In addition, providers may need to consider adapting communication methods with patients during their transplant stay, given potential attention and WM issues within this population.

Categories: Cancer Keyword 1: oncology Keyword 2: attention Keyword 3: working memory **Correspondence:** Hannah-Lise Schofield, Ph.D., ABPP-CN; Children's Hospital of Philadelphia/Perelman School of Medicine at the University of Pennsylvania; schofieldh@chop.edu

48 Longitudinal Language Outcomes in Pediatric Brain Tumor Patients Diagnosed in Early Childhood

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Objective: Pediatric brain tumor (PBT) survivors are at risk for speech (e.g., articulation, prosody, fluency) and language (e.g., vocabulary, grammar, narratives, pragmatics) difficulties (Hodges et al., 2020). It is important to understand what treatment and/or demographic factors are associated with language functioning soon after diagnosis, and what factors are associated with language functioning years after treatment completion. This study characterizes longitudinal language functioning for clinically referred PBT survivors diagnosed in early childhood.

Participants and Methods: Participants were 48 PBT patients (54% supratentorial, 6% disseminated), 21% with NF-1, who were diagnosed by age 6 (M = 43.2 months, SD 24.5) and received tumor-directed intervention including surgery (85%), chemotherapy (69%), and/or radiation therapy (50%). Hearing concerns existed for 29% of the patients. Age at first neuropsychological evaluation was 2-15 years (M=7.6, SD=3.63), age at second neuropsychological evaluation was 5-19 years (M=12.04, SD=3.86), with an average of 4.42 years (SD=2.37) between evaluations. Patients were 63% male, 77% White, 94% non-Hispanic, and fluent English speakers. Verbal IQ, working memory, fluencies, comprehension, memory, and parent-reported functional communication outcomes were assessed as part of comprehensive batteries. Rates of weak performance (1 SD<Mean) were compared to the expected base rate of 16%.

Results: Group means significantly diverged from age-expected performance by the second evaluation in all domains except semantic fluency. Weakness was identified on at least 1 verbal subtest for 79% of the sample at the first evaluation, and for 85% of the sample at the second evaluation. As a group, patients showed a significant increase in the number of weaknesses identified on performance-based measures from the first to second evaluation [t(47) = -3.60, p < .001]. Over half of the sample showed an increase in the rate of verbal weaknesses identified (56.3%). Those with more weaknesses over time had lower IQ at the initial evaluation [t(36) =-2.61, p=.013]. An increase in the number of weaknesses from first to second evaluation was not associated with tumor type/location, treatment modality, or demographic variables.

Conclusions: Brain tumor diagnosis in early childhood during rapid language development is associated with language impairments soon after diagnosis, and years after treatment completion. Causes for continued and increased impairment are multifactorial and risk cannot clearly be identified by demographic and treatment variables alone. Any early language weakness identification should signal need for intervention as the causes for difficulty are complex and these weaknesses are likely to persist and increase over time.

Categories: Cancer Keyword 1: pediatric neuropsychology Keyword 2: brain tumor Keyword 3: language Correspondence: Iris Paltin, PhD, ABPP-CN University of Philadelphia/The Children's Hospital of Philadelphia Paltini@chop.edu

49 Case Study: Cognitive Deficits Associated with Norrie Disease

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Objective: Norrie disease is a rare, x-linked recessive genetic disorder associated with an NDP gene mutation. Males are predominantly affected. Typical symptoms include vision loss

around the time of birth and progressive hearing loss. Cognitive and behavioral abnormalities also occur in 30-50% of individuals, including developmental delays, intellectual disability, cognitive regression, psychosis, and aggression. There is limited research, however, examining the neuropsychological deficits in adulthood resulting from Norrie disease, especially with neuropsychological data and in individuals without other neurological manifestations of the disease, such as seizures. Here, we present the neurocognitive profile of a patient with Norrie disease who presented for a cognitive evaluation in adulthood due to report of more recent memory decline.

Participants and Methods: Mr. Smith is a Caucasian male in his mid-40's who previously underwent genetic testing and was subsequently diagnosed with Norrie disease. As a result of his diagnosis, he experienced complete vision loss since birth and bilateral hearing loss that began in childhood and gradually worsened in adolescence. Medical history was otherwise unremarkable. Developmental milestones were met on time. Historical intelligence testing conducted in elementary school revealed borderline on one intelligence test to high average performance on other intelligence tests. However, he was retained grades several times due to factors such as behavioral disruptions and academic difficulties. He had been employed as an assembly line worker for many years, but had not worked for 10 years prior to the neuropsychological evaluation. Emotionally, he had a longstanding history of anxiety and endorsed mild anxiety and depression at the time of the evaluation. The patient first noticed memory difficulties in adolescence then noticed further decline four years prior to the neuropsychological evaluation (around when he received a left-ear cochlear implant), which had remained stable since onset.

Results: In the context of low average premorbid intellectual functioning, Mr. Smith's neurocognitive profile was notable for difficulties with alphanumeric set-shifting and abstract thinking, with otherwise preserved cognitive functioning. Weaknesses observed on testing may have represented longstanding weaknesses and did not rise to the level of a cognitive disorder. Affective distress was also suspected to have accounted for some of the cognitive lapses the patient reported experiencing with day-to-day functioning. **Conclusions:** The current poster aims to contribute to the limited body of literature