



Neuro UpdateSM

NO LAUGHING MATTER: THE NEUROPSYCHOLOGICAL PROFILE OF A CHILD DIAGNOSED WITH HYPOTHALAMIC HAMARTOMA AND ACCOMPANYING GELASTIC SEIZURES

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Inconsistency has been the most consistent finding in the few research studies focused on cognitive functioning in children with hypothalamic hamartomas. A hypothalamic hamartoma is a rare, benign brain tumor commonly associated with gelastic (i.e., laughing) seizures. Although impairments in cognitive, affective and social functioning have been reported in individuals with this condition, there has been a lack of consensus related to the specific neuropsychological domains affected. Even fewer studies have examined the neurocognitive functioning of children with hypothalamic hamartomas, thus impeding the development of a neuropsychological profile for this population.

PREVIOUS RESEARCH

In an attempt to characterize the cognitive functioning of individuals with hypothalamic hamartomas, Quiske et al. (2006) evaluated 13 juvenile and young adult patients using a standardized neuropsychological battery. Comparisons of individual test performances revealed variable performances across a wide range of cognitive domains.

Impairments most commonly were noted in the intellectual, verbal and nonverbal memory and working memory domains, while visuospatial rotation skills were found to be the most robust. Results from Prigatano et al.'s (2004) study evaluating six children with hypothalamic

hamartomas indicated significant difficulties with speech and language tasks. Deficits also were noted with learning and memory measures, while the visuospatial domain was found to be intact. Commensurate with these findings, results from Frattali et al.'s (2001) study of 13 children with hypothalamic hamartomas demonstrated variable abilities across most neurocognitive measures, with a relative strength emerging on visual processing tasks.

Given both the erratic functioning observed among individuals with hypothalamic hamartomas and the paucity of research examining the neurocognitive functioning of children with this neurological disorder, research

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in this area is clearly warranted. By examining and comparing the functional capacities displayed by children and adolescents with hypothalamic hamartomas and gelastic seizures, it may be possible to identify a pattern of general strengths and weaknesses commonly seen among children with this condition. The following case is presented as the first in a pilot study aimed at developing a more elucidative profile of neurocognitive functioning in this pediatric population.

CASE REPORT

A 7-year-old male had a medical history indicating a diagnosis of hypothalamic hamartoma and accompanying gelastic seizures by age 2. For one month, he was assessed at the video electroencephalogram (vEEG) unit at the Children's Healthcare of Atlanta Epilepsy Center as a part of his presurgical evaluation. Birth history was significant because of a premature delivery, 32 weeks, and associated breathing struggles. Birth weight was 6 pounds, 2 ounces, and developmental milestones were achieved within normal limits. At 24 months, the child experienced his first seizure, with a subsequent seizure one week later.

Results of a magnetic resonance imaging (MRI) test indicated a hypothalamic hamartoma with accompanying gelastic seizures (Figures 1 to 3). Specifically, the MRI revealed a focal mass in the tuber cinereum measuring approximately 1 cm in each plane. Serial MRIs have been stable in comparison. Seizures remain intractable despite a medication regime of Trileptal®. At the time of evaluation, the child was experiencing approximately four to five gelastic seizures per day, each lasting for six to seven seconds. Also of note, the child carried a previous diagnosis of attention-deficit/hyperactivity disorder (ADHD), inattentive type, for which he was taking methylphenidate (Metadate CD).

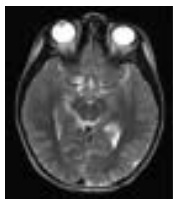


Figure 1 Dorsal view



Figure 2 Posterior view

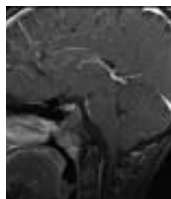


Figure 3 Lateral view

METHODS

The patient was administered neuropsychological tests evaluating his capacities in the following domains: intelligence, e.g., Wechsler Intelligence Scale for Children-Fourth Edition (WISC-IV), academic achievement, e.g., Woodcock Johnson Test of Achievement-Third Edition (WJ-III), receptive and expressive language, e.g., Oral and Written Language Scales (OWLS), visual and motor memory, e.g., Wide Range Assessment of Memory and Learning-Second Edition (WRAML-2), visual perception, e.g., Hooper Visual Organization Test (VOT), and executive functioning, e.g., Drexel Tower of London-Second Edition (TOL). Additionally, several parent-rating scales were used to assess the patient's adaptive skills, e.g., Adaptive Behavior Assessment System-Second Edition (ABAS-2), emotional and behavioral, e.g., Behavior Assessment System for Children-Second Edition (BASC-2) and executive functioning, e.g., Behavior Rating Inventory of Executive Function (BRIEF).

RESULTS

A consistent theme was observed throughout the evaluation; the patient's verbal skills were significantly more robust than his nonverbal abilities (Table 1). Regarding intellectual ability, the child demonstrated low-average verbal capacities in the face of mildly impaired nonverbal skills.

Results from immediate memory measures revealed a similar pattern, as seen in the low-average and borderline abilities that emerged from verbal and visual memory tests respectively. Regarding the standardized measures of executive function, the patient consistently demonstrated more robust abilities with verbal, rather than nonverbal, tasks. Within the nonverbal domain, deficits were noted on tests evaluating tactile perception, e.g., nondominant, left hand only, fine motor dexterity, e.g., bilaterally, mental rotation, visuospatial perception and visuomotor integration.

In contrast, dominant hand dexterity, fine motor speed and visuomotor precision were intact. The patient's performances on measures of short-term attention were more problematic, regardless of the verbal or nonverbal nature of the task, with mild impairment noted in these areas. In keeping with these findings, attentional difficulties also were qualitatively observed throughout the assessment. Additionally, although the patient's mother did not raise concerns related to attentional or executive skills, she reported at-risk concerns related to anxiety, atypical behavior and social functioning.

Measure	SS	Percentile	Measure	SS	Percentile
WISC-IV full scale IQ	64	1	WRAML-2 general memory	77	6
Verbal comprehension	85	16	Verbal memory	77	6
Perceptual reasoning	65	1	Design memory	85	16
Working memory	52	<1	Finger windows	75	5
Processing speed	78	7	Number-letter	65	1
Grooved pegboard	<25	<1	VMI	70	2
Hooper VOT	51	<1	Drexel TOL-2	<60	<1

SS=Standard score (mean=100, SD±15)

DISCUSSION

Overall, the patient evaluated for this study evidenced many of the cognitive deficits reported in previous studies of children with hypothalamic hamartomas and gelastic seizures, e.g., compromised intellectual abilities, impairments in attentional and memory skills and deficits in social functioning. Consistent with results from studies by Quiske et al. (2006), Prigatano et al. (2001) and Frattali et al. (2001), the findings from the current study highlight the adverse effects of hypothalamic hamartomas and gelastic seizures on the neuropsychological functioning of children and adolescents. Specifically, overall intellectual functioning and learning and memory abilities often are compromised among this population.

Also of note, the contradictory findings that emerged in other functional domains. Unlike the intact performances described by Quiske et al. (2006) on measures of mental rotation, the current study participant demonstrated a significant weakness in this domain, performing in the moderately impaired range. The patient also struggled with measures of visual perception, which is discrepant with results from both Prigatano et al.'s (2001) and Frattali et al.'s (2001) studies.

Further, the patient did not evidence the affective regulation difficulties commonly observed in individuals with hypothalamic hamartomas. Thus, while patients with hypothalamic hamartomas are seemingly prone to impairment in both the cognitive and memory domains, the effects of this condition on other areas of neurocognitive function remain unclear. Consequently, continued research examining the neuropsychological functioning of children with hypothalamic hamartomas and accompanying gelastic seizures is warranted.

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