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PSYCHIATRIC DIAGNOSES IN HYPOTHALAMIC HAMARTOMA PATIENTS

COMMENTS

Since initating the hypothalamic hamartoma consortium at our institution in 2003, we have operated and treated more than 100 patients with this condition. Barrow has now acquired the largest institutional experience with the surgical treatment of these rare developmental abnormalities. Overall, improvements in surgical technique have improved outcomes, eradicating or ameliorating the intractable epilepsy too often associated with these lesions.

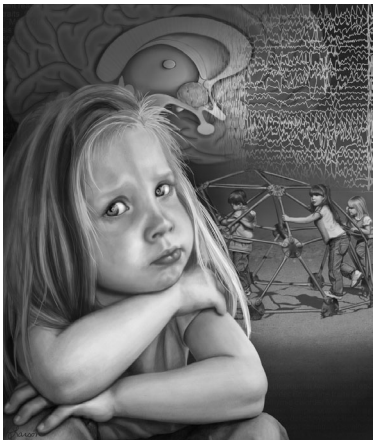
Nonetheless, the behavioral and psychiatric disorders associated with hypothalamic hamartomas can be as devastating to patients and their families as the seizures. Not all patients may regain a normal level of function. As of yet, however, no neuropsychological tool is available for the assessment of children and adults with a hypothalamic hamartoma. Despite efforts in the neuropsychological community to relate these patients' psychiatric and behavioral disorders to their seizure history, no significant relationship has been found. The lack of this information impedes the development of neuropsychological treatments or rehabilitation best suited to help patients and families.

Dr. Prigatano, Director of the Division of Neuropsychology, and colleagues are therefore pursuing this under-researched aspect of hypothalamic hamartomas. In this issue, they report the results of a preliminary study documenting the variability of the diagnostic terms used to describe 57 children and adults with hypothalamic hamartomas and refractory epilepsy. Their goal was to explore the variability of diagnostic terms applied to these patients by previous clinicians to identify any relationships between these patients' psychiatric and behavioral diagnoses and estimates of their intellectual functioning. As this group notes, systematic neuropsychological studies are needed help explain the various psychiatric disturbances seen in patients with this unfortunate condition.

Also in this issue, Aliabadi et al. review a case that will alert clinicians to the diagnostic similarities between diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis and how to distinguish between the overlapping clinical features associated with these two conditions. Despite the clinical similarities of these two pathologies, their treatments differ considerably. Consequently, an early and accurate diagnosis is crucial to the successful management of patients with severe cervical ankylosis.

In another article, Wait and coworkers present clinical images from a patient originally treated for symptoms of tectal compression and intermittent hydrocephalus. Postoperatively, the patient developed a de novo simple cerebellar cyst. This entity was new to our institution and does not appear to have been reported previously. Finally, readers will find helpful indices of all the articles published in 2005 and 2006. We hope our readers find these articles interesting and out of the ordinary. As always, we are pleased to share our clinical findings with students of the neurosciences. However, we need your help too. Please consider using the enclosed self-addressed and stamped envelope to forward a tax-deductible donation that will help us continue to provide this journal free of charge. Thank you for your help.

Robert F. Spetzler, MD
Editor-in-Chief



This issue's cover illustrates a child with emotional and social difficulties associated with a hypothalamic hamartoma. This benign tumor, which involves the hypothalamic nuclei and causes gelastic seizures, can lead to various behavioral, psychiatric, and cognitive disorders. See the article by Prigatano et al. on page 4. The illustration is by Kristen Larson.