

R, Nashef L, et al. The clinical spectrum of epilepsy in children and adults with hypothalamic hamartoma. Epilepsia October 2003;44:1310-1319). (Reprints: Dr N Mullatti, Department of Clinical Neurophysiology, Kings College Hospital, Denmark Hill, Lonsdon SE5 9RS, UK).

COMMENT. Gelastic seizures are the rule with hypothalamic hamartoma (HH) and early-onset epilepsy, but they frequently evolve into other and milder types of epilepsy in adolescence and adulthood. In adult-onset epilepsy with HH, gelastic seizures are less frequent and milder, and epilepsy is often of the complex partial, tonic, or atypical absence types. Cognitive and behavioral disorders are less frequent in adult-onset cases of epilepsy with HH.

## SEIZURE DISORDERS

### **HIPPOCAMPAL ABNORMALITIES AFTER PROLONGED FEBRILE CONVULSIONS**

Hippocampal volume and T2 relaxation times were determined in an MRI study of 14 children with prolonged febrile convulsions (PFC) who were investigated, 1) within 5 days of a PFC, and 2) at follow-up 4-8 months after the acute study, at the Institute of Child Health, University College, and Great Ormond Street Hospital, London, UK. The initial study previously published (Scott et al, 2002) showed that children examined by MRI within 48 h of a PFC had large hippocampal volumes and prolongation of T2 relaxation times, whereas those with MRI delayed >48 h but within 5 days of PFC had large hippocampal volumes and normal T2 relaxation time. The data were suggestive of hippocampal edema that is resolving within 5 days of a PFC.

Of 14 patients examined at 4-8 month follow-up, 4 had suffered further seizures: 2 had short FC, 1 had PFC and 1 had non-febrile seizures. Repeat MRI showed a significant reduction in hippocampal volume and T2 relaxation time compared to the first exam, and volumes were not different from a control population. In addition, hippocampal volume asymmetry was significantly increased at follow-up compared to initial data, findings consistent with injury and neuronal loss associated with PC. It is postulated that some selectively vulnerable patients may develop mesial temporal sclerosis after a lag period of several years following the PFC or alternatively, the hippocampal asymmetry may represent a post-acute edema return to a pre-existing hippocampal abnormality that antedates the PFC. (Scott RC, King MD, Gadian DG, Neville BGR, Connelly A. Hippocampal abnormalities after prolonged febrile convulsion: a longitudinal MRI study. Brain November 2003;126:2551-2557). (Respond: Dr Rod C Scott, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. In patients with intractable complex partial seizures, those with mesial temporal sclerosis (MTS) had a history of PFC in 33% whereas those without MTS had PFC in only 7% (Falconer, MA, 1972, 1976; see Nelson and Ellenberg, 1981). A causative association between prolonged or complex FC and hippocampal injury and MTS is suggested by clinical and animal studies, but a pre-existing subtle hippocampal maldevelopment is possible in some cases of complex partial epilepsy.

**Magnetic brain source imaging (MSI) of focal epileptic activity** is reported in 455 cases of epilepsy examined preoperatively at University of Erlangen-Nuernberg, Germany (Stefan H et al. *Brain* 2003;126:2396-2405). The average sensitivity of magnetoencephalography (MEG) for specific epileptic activity was 70%, whereas MSI identified the lobe to be resected in 89% of 131 patients who underwent surgery. Those with extratemporal epilepsies were identified in a higher percentage than temporal lobe cases. In a study of 20 patients with **idiopathic generalized epilepsy** compared to healthy controls, magnetic resonance spectroscopic imaging showed a progressive thalamic neuronal dysfunction, independent of the amount of spike and wave activity (Bernasconi A et al. *Brain* 2003;126:2447-2454). The complementary use of MEG and EEG was useful in localizing the origin of cortical myoclonus in the right temporo-occipital cortex in a 15 year-old girl with Lafora-body disease (Verrotti A et al. *Acta Paediatr* Oct 2003;92:1218-1222).

## **HEMISPHERECTOMY FOR INTRACTABLE EPILEPSY**

Charts of 111 patients, ages 2 months to 20 years, who underwent hemidecortication for intractable unihemispheric epilepsy in the period 1975-2001 were reviewed at Johns Hopkins Hospital. The average time from seizure onset to operation was 3.6 years. Follow-up ranged from 3 months to 22 years (3 died in the perioperative period, 2 died later due to seizures, and 3 were lost to follow-up). Seizures were controlled in 65%, 21% had occasional seizures, and 14% were not benefited. One or no anticonvulsant was continued in 80%, and 89% were ambulatory. Outcome was correlated with etiology; patients with seizures caused by migrational disorders were controlled in 51%, whereas those with other seizure etiologies (Rasmussen and congenital vascular) were seizure-free in 71% ( $p=0.05$ ). (Kossoff EH, Vining EPG, Pillas DJ et al. Hemispherectomy for intractable unihemispheric epilepsy. Etiology vs outcome. *Neurology* October (1 of 2) 2003;61:887-890). (Reprints: Dr Eric Kossoff, Jefferson 128, The Johns Hopkins Hospital, 600 North Wolfe St, Baltimore, MD 21287).

COMMENT. The Johns Hopkins extensive series of hemispherectomies for intractable seizures demonstrates a beneficial response in 86% of cases, and the outcome is particularly favorable in patients with seizures due to Rasmussen syndrome and vascular disorders. The ideal time to operate remains undetermined. Significant morbidity or mortality occurred in 7%.

## **RASMUSSEN SYNDROME: RESPONSE TO THALIDOMIDE**

A 13-year-old girl with Rasmussen syndrome diagnosed at 5 years, and right-sided seizures refractory to various therapies, including hemispherectomy, responded to thalidomide and is reported from Belgrade, Yugoslavia. Initial control with carbamazepine, valproate and clonazepam was followed by relapse and a more extensive right hemiparesis. Despite resection of the cortical epileptogenic zones in the left hemisphere at 9 years of age, and trials of different anticonvulsants, acyclovir, g-globulin, iv methylprednisolone, oral prednisone, and plasma exchange, seizure frequency