diagnosis of supratentorial tumors was delayed for an average of 2 years, whereas infratentorial tumors were diagnosed within 3 months of the initial seizure (Backus RE, Millichap JG. Pediatrics 1962;29:978-984).

SEIZURE DISORDERS

PROGNOSIS OF INFANTILE SPASMS AND L-G SYNDROME

The occurrence, outcome, and prognostic factors of infantile spasms (IS) and Lennox-Gastaut syndrome (LGS) were determined in children treated in the Department of Pediatrics, University of Oulu, Finland, from Jan 1976 to Dec 1993. Thirty seven had IS (0.41/1000 live births) and 25 had LGS (0.28/1000 live births). Ten (27%) of the patients with IS who later developed LGS (40% of LGS cases) had symptomatic epilepsy, were mentally retarded, and their seizures were uncontrolled at 10 year follow-up. Symptomatic epilepsy (30 (81%) IS and 17 (68%) LGS) had congenital or genetic etiologies in almost all cases (87% of IS, 100% of LGS). Cryptogenic epilepsy in 7 (19%) of the IS cases had a favorable prognosis, whereas in 8 (32%) of LGS cases, a cryptogenic etiology did not decrease the risk for a poor outcome. The majority received ACTH and polytherapy. (Rantala H, Putkonen T. Occurrence, outcome, and prognostic factors of infantile spasms and Lennox-Gastaut syndrome. Epilepsia March 1999;40:286-289). (Reprints: Dr H Rantala, Department of Pediatrics, University of Oulu, FIN90220 Oulu 22, Finland).

COMMENT. The prevalence of infantile spasms (IS) in a primary university pediatric population in Finland is 0.4/1000 live births and that of Lennox-Gastaut syndrome (LGS) is similar. IS evolves into LGS in 27% of cases and these are symptomatic epilepsies, with a poor prognosis. Cryptogenic etiology has a favorable prognosis for IS but not in LGS cases.

Vigabatrin in the treatment of infantile spasms has been studied retrospectively in 25 infants (19 symptomatic, 6 cryptogenic cases) followed at the Children's Hospital of Michigan, Detroit, MI. (Koo B. Pediatr Neurol Feb 1999;20:106-110). Clinical improvement was obtained in 16 (64%), and EEG improvement in 17 (68%). EEG and cognitive decline and/or more frequent spasms occurred in 7 (28%), often associated with larger VGB doses (>100 mg/kg daily). Smaller doses and EEG monitoring are recommended, since EEG and cognitive deterioration may occur despite clinical control of spasms.

Efficacy of Lamotrigine in refractory neonatal seizures of unknown etiology is reported in a single newborn treated at the Royal Alexandra Hospital for Children, Parramutta, NSW, Australia. (Barr PA, Buettiker VE, Antony JH. Pediatr Neurol Feb 1999;20:161-163). The EEG showed a burst-suppression pattern, and seizures were mainly generalized. Conventional AEDs were ineffective, vigabatrin (105 mg/kg/d) was partially effective, and the addition of lamotrigine (4.4 mg/kg/d) was followed by a sustained seizure control.

GELASTIC EPILEPSY AND HYPOTHALAMIC HAMARTOMA

The causes, clinical manifestations, and evolution of gelastic seizures (GS) were studied, using video-EEG and MRI, in 9 patients observed between 1986 and 1997 at the Epilepsy Center, Federico II University, Naples, Italy. Seizures were frequent (several/day) and characterized by laughing attacks, sometimes with facial flushing, and rarely with loss of contact. Age at onset was less than a year in 3, and < 12 years in 8. All older patients reported feelings of embarrassment,

and attempts to conceal the seizure (eg. by feigning a sneeze); no feeling of amusement accompanied the laughter. Of 5 patients with symptomatic, localization-related epilepsy, 4 had hypothalamic hamartoma (HH) and 1 had tuberous sclerosis (TS). In 4 patients with cryptogenic gelastic seizures, the MRI was negative but seizures and EEG showed focal characteristics. Seizures were drug resistant in the HH cases, the TS patient responded to vigabatrin, and cryptogenic GS showed a variable outcome with partial seizure control. Two HH cases (ages 3 yrs and 28 yrs) showed complete or partial benefit following total surgical removal. Precocious puberty and cognitive deterioration, well recognized complications of the gelastic epilepsy-hypothalamic hamartoma syndrome of childhood, were not encountered in this series of patients. Cryptogenic cases of GS were usually more benign than the symptomatic HH cases. (Striano S, Meo R, Bilo L et al. Gelastic epilepsy: Symptomatic and crytogenic cases. Epilepsia March 1999;40:294-302). (Reprints: Prof Salvatore Striano, Viale dei Pini 101, 80131 Naples, Italy).

COMMENT. The childhood epileptic syndrome of early-onset gelastic seizures (GS; from the Greek *'gelos'* meaning mirth), hypothalamic hamartoma (HH), and precocious puberty (PP) is relatively rare but important in diagnosis and management. Frequent recurrent attacks of laughter, especially when accompanied by facial flushing, in a young infant or child may represent a recognized form of epilepsy and should prompt an MRI and EEG. The prognosis is usually poor, the GS evolving into symptomatic generalized epilepsy and leading to cognitive deterioration, but early recognition and surgical ablation of the hamartoma may occasionally result in control of drug refractory seizures and prevention of mental retardation.

The authors in this relatively large series of cases of gelastic epilepsy, including GS that are symptomatic of hamartomas and an equal number of cryptogenic cases, allude to the absence of precocious puberty, usually considered a classic sign of the syndrome, a later onset of seizures in some, and the relatively benign outcome of non-hamartoma cases.

Previous reports of gelastic epilepsy have usually involved only isolated cases. In 3 cases studied at Johns Hopkins University, GS originated in the anterior cingulate or fusiform gyri, and seizures were controlled following surgical removal of a cavernous hemangioma in one (Arroya S et al. Brain 1993;116:757-780). Infantile spasms are often associated with attacks of paroxysmal laughter (Lacy JR, Penry JK. Infantile Spasms. New York, Raven Press, 1976). The classic epilepsy monographs of Turner WA (1907) and Lennox WG (1960;pp280-1) have references to compulsive laughter as a manifestation of epilepsy. Reference to GS is surprisingly absent from Penfield's monograph (1954). (See Progress in Pediatric Neurology II, Chicago, PNB Publishers, 1994;pp41-42, for reports and commentary on gelastic epilepsy).

LONG-TERM OUTCOME OF LANDAU-KLEFFNER SYNDROME

The types of language disorders in 4 young adults with Landau-Kleffner syndrome were analyzed at long-term follow-up at the National Institute of Mental Health, National Center of Neurology and Psychiatry, Chiba, Japan. Age of onset was 4 to 6 years, and the first symptoms of language disorder were an auditory verbal agnosia in 3 cases, and sensory aphasia in 1. All patients showed a sequence of deterioration followed by recovery of language function without intellectual handicap. Sequelae included disability in spoken language, auditory verbal and music perception, and lower Wechsler Verbal scores compared to Performance IQ scores. Epilepsy began at 5 years in 2, and EEG epileptiform abnormalities occurred in all patients. CTs were normal in 3 and one showed an