

with numerous previous reports of similar clinical febrile seizure studies. Between 1924 and 1965, 51 articles involving approximately 10,000 febrile seizure patients were published in the world literature. (Millichap JG. Febrile Convulsions, New York, Macmillan, 1968). A male preponderance was established in 29 series, with a mean sex ratio of 1.4 to 1, a family history of febrile seizures was found in 30%, and the mean threshold convulsive temperature was 104.0°F. In contrast to the Seattle findings, otitis media accounted for only 2.9% of associated fevers, and tonsillitis or pharyngitis was by far the most frequent illness, occurring in 59% of febrile episodes. Focal seizures were reported in a mean of 11%, and 14% in one prospective study. The incidence of Todd's paresis was 3.7% and similar to that observed in Seattle. Of prognostic importance is the confirmation of age at onset (< 1 year) as a risk factor for complex febrile seizures in this study.

### EPILEPSY WITH MYOCLONIC ABSENCES

Resistance to therapy and learning disabilities are stressed as frequent complications of the syndrome of "epilepsy with myoclonic absences" reported in 8 children, ages 6 to 16 years, seen at the University Hospital of Wales in a 10 year period. The mean age at onset was 4.9 years. Febrile seizures had occurred in siblings of 3 patients. Myoclonic absences were brief, and they could be precipitated by hyperventilation. Loss of awareness was associated with bilateral jerking of the head and upper limbs, and the EEG showed rhythmic 3 c/s spike-wave discharges. The majority had generalized tonic-clonic or atstatic seizures and cerebellar ataxia in addition. All became learning disabled and 7 had behavioral problems, including restlessness and impulsiveness. Treatment with lamotrigine and valproate was partially effective. (Manonmani V, Wallace SJ. Epilepsy with myoclonic absences. Arch Dis Child April 1994;70:288-290). (Respond: Dr Wallace, Dept Paediatric Neurology, University Hospital of Wales, Heath Park, Cardiff CF4 4XW, Wales, UK).

COMMENT. The response to lamotrigine (LTG) reported by Wallace in this and a previous report is in contrast to the experience of Schlumberger E et al at the Hopital Saint Vincent de Paul, Paris, France. (Epilepsia March/April 1994;35:359-367). Of 9 patients with myoclonic absence epilepsy treated with LTG, none was seizure-free at 3 months, 4 improved and 5 were unchanged. Side effects included skin rash, especially when LTG was added to VPA therapy, ataxia, drowsiness, and vomiting. The differentiation of this syndrome from typical absence epilepsy is important because of the poor response to treatment and the unfavorable long term outcome.

### GENETICS OF EARLY CHILDHOOD ABSENCE EPILEPSY

The clinical and EEG family data of 140 cases of early childhood epilepsy with absences selected from the epilepsy family archive are reported from the Neuropaediatric Department of the University of Kiel, Germany. Patients with absences manifesting between the 1st and 5th year of age were selected for study. Two groups were formed: 1) GTCS, those presenting with generalized tonic-clonic seizures (90 cases); and 2) non-GTCS, presenting with absences (50 cases). GTCS at onset were afebrile or febrile. In 43% of probands absence were combined with myoclonic and/or myoclonic atstatic seizures. Parents and their sibs of group 1 had seizures twice as often as parents and sibs in group 2. The

EEG of relatives showed elevated incidences of spike and wave and photosensitivity in both groups. However, in parents of the non-GTCS group, EEG abnormality was more frequent than in parents of the GTCS group. Mothers' EEG was the best predictor of seizure risk in probands' siblings. Early childhood epilepsy with absences overlaps with early onset GTCS and myoclonic astatic epilepsy on one hand and with childhood absence epilepsy on the other. This syndrome could not be regarded as a specific entity. The analysis supports the assumption of heterogeneity within early childhood absence epilepsy. (Dooze H. Absence epilepsy of early childhood - genetic aspects. Eur J Pediatr May 1994;153:372-377). (Respond: Dr H Dooze, Norddeutsches Epilepsie-Zentrum, D-24223 Ralsdorf, Germany).

COMMENT. Three type of absence epilepsy are distinguished by the International Classification: 1) childhood absence with absence as presenting symptom, 2) juvenile absence with or without GTCS, and 3) epilepsy with myoclonic absences. Absence epilepsy of early childhood has not been distinguished as a specific entity, although Dooze has demonstrated some characteristics which might allow differentiation from childhood absence. These include: onset before 5 years of age with absences or GTCS, male preponderance, associated myoclonic and/or myoclonic astatic seizures, poor response to AEDs, and unfavorable psychologic and social development in children with GTCS. The clinical and family data reported here are considered to support genetic heterogeneity within the disorder. Both non-GTCS and GTCS forms of the syndrome appear to be part of the larger spectrum of idiopathic generalized epilepsies of childhood.

## GABA SYNTHESIS AND PYRIDOXINE SEIZURES

A reduction in pyridoxal-5-phosphate (PLP) dependent enzyme, glutamic acid decarboxylase (GAD), which synthesizes GABA, is reported in a 3 month-old infant with seizures responsive to pyridoxine treated at University of California, Davis. The infant had asynchronous jerking of arms and legs and lip smacking at birth which responded to phenobarbital and phenytoin. MRI showed enlarged ventricles, and the EEG demonstrated bitemporal and left frontal epileptiform activity. Seizures recurred at 7 weeks and were not responsive to phenobarbital and carbamazepine. When admitted at 3 months, 100 mg pyridoxine IV stopped a seizure within 5 minutes. Anticonvulsants were discontinued and electrographic and clinical seizure activity was controlled with oral pyridoxine 25 mg daily. PLP independent GAD activities measured in skin fibroblasts of the patient and 5 controls were similar, whereas the patient's PLP dependent GAD activity was reduced. (Gospe SM Jr, et al. Reduced GABA synthesis in pyridoxine-dependent seizures. Lancet May 7 1994;343:1133-34). (Respond: Dr Sidney M Gospe Jr, Division of Child Neurology, University of California, Davis Medical Center, 2315 Stockton Boulevard, Sacramento, CA 95817, USA).

COMMENT. An alteration in the function of PLP dependent GAD appears to be responsible for the autosomal recessive syndrome of pyridoxine dependent epilepsy.