and the consequent risk of a heightened prevalence of febrile seizure recurrence emphasizes the importance of parental education in the management of fever and febrile seizures. In the absence of a safe and satisfactory alternative to phenobarbital, parental anxiety may be allayed by the prescription of intermittent prophylactic treatment with diazepam at times of fevers but poor compliance minimizes its effectiveness in practice. The home use of rectal diazepam, employed more frequently in Europe than in the U.S., may offer an alternative method of parental involvement in selected cases.

HYPEREXPLEXIA OR HEREDITARY STIFF BABY SYNDROME

The stiff baby syndrome and its diagnostic distinction from epilepsy is reviewed from the Service de Neuronatologie, Pavillon de la Mere et l'Enfant, Nantes, France. The disease hyperexplexia was first described in 1962 by Kok and Bruyn in 29 members of one family and occurred as a dominant autosomal transmission. It was distinguished by a permanent hypertonia that is heightened by the slightest stimulus. The hypertonia was noted at birth and became less pronounced during the first year of life but later could lead to repeated falls. The electromyogram showed persistent activity even at rest and the activity was abolished by diazepam. Lingam S, Wilson J, and Hart E named the condition "hereditary stiff baby syndrome" (Am J Dis Child 1981; 135:909). The child has a fixed stare and an expression of anxiety. The hypertonia diminishes during sleep and increases with the slightest psychic or tactile stimulus. Attacks of hypertonia may involve respiratory muscles and lead to apneas which can endanger the child's life. Digestive disorders including vomiting are usually associated with a hiatal hernia. The electroencephalogram is normal. (Tohier C et al. Hyperexplexia or stiff baby syndrome. Arch Dis Child April 1991; 66:460-461).

COMMENT. In addition to myoclonic epilepsy, the differential diagnosis includes the stiff man syndrome which is not hereditary but which may occur in children (Millichap JG,unpublished observation), the Isaacs-Mertens syndrome with distal hypertonia and fasciculations, the jumping Frenchmen of Maine syndrome with violent starts induced by slight stimuli and associated with echolalia and echopraxia, and Gilles de la Tourette syndrome. The pathology of stiff man syndrome has been localized to the spinal interneurons but the mechanism of hyperexplexia is controversial. Treatment with diazepam is effective.

POLYPHARMACOTHERAPY IN INSTITUTIONALIZED EPILEPTIC CHILDREN

An attempt to minimize polypharmacotherapy, to discontinue the use of phenobarbital, and to assess the relation between drug levels and antiepileptic effect in institutionalized severely retarded children is reported from the Department of Pediatrics, St. Goran's Hospital, Stockholm, and the Division of Clinical Pharmacology, University Hospital, Uppsala, Sweden. Nine severely mentally retarded patients