

epilepticus in children; it is investigative and nonapproved for use in neonates. Prospective studies may be justified.

In an Editorial, Sankar R and Painter MJ (**Neurology** 2005;64:776-777) applaud the study of midazolam and the promising results. However, they raise concerns about potential neurotoxicity of GABA agonists in the immature brain. AMPA antagonists, such as topiramate, might be a safer class of drug to promote in IV form for trial in neonates.

BENIGN FAMILIAL INFANTILE SEIZURES

The history, classification, clinical and EEG features, genetics, differential diagnosis, and outcome of “benign familial infantile seizures” (BFIS) are reviewed from the Neurology Department, Bambino Gesù Children Hospital, Rome, Italy. First reported by Fukuyama (1963) as partial seizures occurring in the first 2 years, idiopathic, and with a benign outcome, the syndrome was later described as benign partial epilepsy of infancy with complex partial seizures (BPE and CPS) and BPE with secondarily generalized seizures in infancy (Watanabe et al. 1987, 1990, and 1993). The term “benign infantile familial convulsions” was proposed by the present author (Vigevano F et al. 1992), cases showing an autosomal dominant inheritance. Finally, in the ILAE classification (2001) the term “benign familial infantile seizures” was preferred, and two forms were listed, familial and nonfamilial. The inheritance is heterogeneous, with chromosome markers on chromosomes 19, 16 (BIFC and paroxysmal choreoathetosis), and on chromosome 2. Two families were also described with onset at the 2nd month, with mutations in the sodium-channel gene SCN2A, and two with neonatal onset had genetic mutations associated with potassium channels. Some cases have been associated with diarrhea and rotavirus.

Clinical characteristics are as follows: Family history of similar seizures; normal early development; onset 3-10 months; normal neurologic exam; seizures in clusters; partial (occipito-parietal) seizures; normal interictal EEG; benign course; and normal developmental outcome. In the absence of a family history of BFIS, early diagnosis may be difficult and only by exclusion of possible etiological factors or by identification of the genetic marker. Sporadic cases may carry the same genetic marker as familial ones, with less expressivity. (Vigevano F. Benign familial infantile seizures. **Brain Dev** April 2005;27:172-177). (Respond: F Vigevano; E-mail: vigevano@opbg.net).

COMMENT. The syndrome of BFIS has characteristic clinical features and benign outcome but variable genetic mutations associated with a channelopathy. The decision to treat depends on the severity and frequency of seizures and the family history. The author cites untreated familial cases having isolated or brief clusters up to 1 year of age.

PROGNOSIS OF BENIGN MYOCLONIC EPILEPSY OF INFANCY

Neuropsychological, cognitive, and behavioral outcome was studied in a long-term follow-up of 7 patients with benign myoclonic epilepsy in infancy (BMEI) at Università di Palermo, Italy. Mean age at onset of myoclonic seizures (MS) was 15 months (range, 7-35 months). Febrile convulsions had occurred before the onset of MS in 3 infants. Myoclonic jerks involved mainly the upper limbs, with nodding and upward gaze deviation in some, and flexing of the body and lower limbs. Ictal EEG recordings showed generalized spike-wave