

Fukuyama Y (**Epilepsia** 2010;51:2216-7), in a commentary from Japan, prefers the term West syndrome to infantile spasms, and notes differences in treatment protocols in US and Japan. In Japan, the dose of ACTH recommended is smaller and the courses, shorter. Vitamin B6 in large doses is advocated as the first-line treatment for newly diagnosed West syndrome patients. VGB is not available in Japan.

Commentaries from Europe and the UK (Dulac O et al. **Epilepsia** 2010;51:2218-9) favor a selective diagnostic and therapeutic approach. Given etiologies for IS determine specific clinical/EEG/imaging patterns. Hypoxic-ischemic encephalopathy, a known cause of WS, has a different course for pre-term and full-term infants. IS following preterm delivery of infants with HIE respond well to treatment, whereas HIE-related spasms following full-term delivery tend to be more severe and refractory. Spasms without hypsarrhythmia, but only focal or multifocal spikes, may suggest focal cortical dysplasia or tuberous sclerosis complex as a cause. These authors are impressed with VGB, as first-line therapy, and consider the ketogenic diet no more effective than pyridoxine or conventional AEDs. Parents should be informed of the importance of the EEG in diagnosis, and pediatricians made aware of the early manifestations of WS before the onset of typical spasms. A lack of visual contact or awareness in an infant may signal the onset of WS and the need for wake and sleep EEG, if apparent visual impairment is unexplained by ophthalmologic examination. Early diagnosis and treatment appear to be the key to success in the management of West syndrome.

LONG-TERM OUTCOME OF ENCEPHALOPATHY WITH STATUS EPILEPTICUS DURING SLEEP SYNDROME (ESESS)

Researchers from the Epilepsy Unit, Helsinki University Central Hospital, Finland, prospectively evaluated the efficacy of antiepileptic drug treatment and long-term (5 years or longer) cognitive outcome in 32 children with ESESS. Epilepsies were atypical rolandic (AR) in 6 children, associated with Landau-Kleffner syndrome (LKS) in 9, and symptomatic in 17. Prospective treatment with valproate (VPA) combined with ethosuximide (ESM) in 17 children was effective in 3 (18%). Electrical status epilepticus during sleep was abolished in 16 patients. Pre-ESESS cognitive level was regained in a total of 10 (31%) children (4 with AR, 3 with LKS, and 3 with symptomatic epilepsy). The majority had permanent cognitive impairment. Younger age at ESESS diagnosis, lower IQ at diagnosis, and no response to drug treatment were predictive of an unfavorable cognitive outcome. Cognitive outcome depends on treatment response, on EEG and seizures, and on etiology. VPA combined with ESM was the most effective drug treatment. Eight of 16 nonresponders underwent epilepsy surgery. (Liukkonen E, Kantola-Sorsa E, Paetau R, Gaily E, Peltola M, Granstrom M-L. Long-term outcome of 32 children with encephalopathy with status epilepticus during sleep, or ESES syndrome. **Epilepsia** Oct 2010;51(10):2023-2032). (Respond: Dr Elina Liukkonen, Epilepsy Unit, Helsinki University Central Hospital, PoB 280, 00029 HUS, Finland. E-mail: elina.liukkonen@hus.fi).

COMMENT. SES (or CSWS), the term used for the EEG phenomenon, is defined as spike and wave discharge in >85% of non-rapid eye movement (REM) sleep. The encephalopathy ESESS has a poor long-term outcome. Only one third cases regain a

normal cognitive level. Control of seizures and abolition of SES are essential for cognitive recovery. Outcome is dependent on etiology. Patients with rolandic epilepsy and ESESS have the best prognosis, whereas those with ESESS caused by LKS or symptomatic epilepsy are usually impaired. Children with congenital hemiplegia, hydrocephalus, or thalamic injury with early onset epilepsy are at increased risk of ESESS, and should be carefully monitored. Surgery should be considered early in treatment of drug-resistant symptomatic ESESS.

In a series of 30 patients with ESES treated 1994-2007 in Tel Aviv, Israel, the syndrome evolved from benign partial epilepsy in 11 (37%), and another third had an underlying structural brain anomaly. The most effective AEDs were levetiracetam and clobazam, whereas valproate and ethosuximide were ineffective. Residual intellectual deficit correlated with duration of ESES. (Kramer U et al. **Epilepsia** 2009;50(6):1517-1524).

INFECTIOUS DISORDERS

LONG-TERM OUTCOMES OF ACUTE ENCEPHALITIS

Researchers at the Astrid Lindgren Children's Hospital and Karolinska Institute, Stockholm, Sweden, reviewed medical records of 71 of 93 children who were treated for acute encephalitis at 5 weeks to 17 years of age in 2000-2004, using questionnaires and a structured telephone interview conducted with the parents. Fifteen children with the most severe symptoms at time of discharge underwent EEG and tests of reaction time and working memory. Mean age at onset of encephalitis was 6.7 y (range 0-17) and time to follow-up evaluation was 5.4 y (range 3-8). Males outnumbered females 47 to 24 or 2:1. The cause was known in 37 (52%). Persisting symptoms were reported in 25 (60%) of 42 children >5 y of age and in 13 (45%) of 29 children <5 y of age. Symptoms resolved completely in 24 (34%) of 71 children (within 6 months in 21 children and by 1 year in 3). Residual symptoms were reported in 17 (40%) of 42 children >5 y of age and in 8 (28%) of 29 children <5 y of age at time of acute illness. The most common residual symptoms in the older group were personality changes, poor memory, noise sensitivity, and poor concentration, and poor concentration and feelings of frustration in the <5 y group. A confirmed/probable microbial cause was established in 52% of the children. Prevalence of persisting symptoms was similar in cases of known and unknown cause, and all agents were equally causative of persisting symptoms, even those considered benign. Factors indicative of a poor prognosis and persisting symptoms at follow-up included admission to ICU in acute phase, and fever, seizures, EEG abnormalities, and moderate/severe symptoms at discharge. Postencephalitic epilepsy developed in 7 (10%) children. Girls had a fivefold increased risk of epilepsy compared with boys (P<0.05). Children with seizures during the acute illness had an eight-fold increased risk of epilepsy (P<0.05). All children who developed epilepsy had abnormal EEG findings during acute encephalitis, and recordings were abnormal in 9 of 15 with repeat EEGs at follow-up. Cognitive testing showed better results with increasing age for reaction time but not for working memory. Children with encephalitis had slower reaction times than controls. (Fowler A, Stodberg T, Eriksson M, Wickstrom R. Long-term outcomes of acute encephalitis in childhood. **Pediatrics** Oct 2010;126:e828-e835). (Respond: Asa