FEBRILE-INFECTION-RELATED EPILEPSY SYNDROME (FIRES)

Researchers at Tel Aviv University, Israel, and other epilepsy centers in Taiwan; Rome, Italy; Boston, USA; Paris, France; Vogtatreuth and Kiel, Germany; and Salzburg, Austria studied retrospectively the pathogenesis, treatment, and outcome of 77 children diagnosed with febrile infection-related epilepsy syndrome (FIRES) and reported in 8 publications between 2001 and 2010. Median age at onset was 8 years (range 3-17 years), and the male-to-female ratio was 4:3. Prolonged refractory status epilepticus or multiple daily seizures (partial or secondarily generalized) were preceded by fever for a median of 4 days and nonspecific infection (upper respiratory or gastrointestinal) in 96% of patients. Mechanical ventilation was required for a median duration of 41 days. CSF showed minimal pleocytosis (57%) and oligoclonal bands (4/12) but the etiology was undetermined. Tests for viral infection were negative. Genetic test for SCN1A mutation was negative in 2 patients examined. Treatment with antiepileptic drugs or steroids was ineffective. Treatments of possible benefit included IVIG (in 2 of 30 patients), ketogenic diet (in 1 of 4), and prolonged barbiturate anesthesia (1 of 46). Nine (12%) patients died during the acute illness; of 68 survivors, 56 (82%) had cognitive disabilities, and 63 (93%) had refractory epilepsy at follow-up. Poor cognitive outcome was significantly associated with younger age at FIRES onset, and higher log of burst-suppression coma duration. Some patients had severe peripheral neuropathy and ataxia as sequelae. There was no correlation between mortality or degree of cognitive impairment and EEG foci (temporal or frontotemporal in 32 [54%] of 59) during the acute phase, MRI abnormalities (signal hyperintensities and later atrophy in hippocampi), and duration of mechanical ventilation. (Kramer U, Chi C-S, Lin K-L, et al. Febrile infection-related epilepsy syndrome (FIRES): pathogenesis, treatment, and outcome. A multicenter study on 77 children. **Epilepsia** November 2011;52(11):1956-1965). (Respond: Uri Kramer MD, Pediatric Epilepsy Service, Tel Aviv Sourasky Medical Center 6 Weitzman St, Tel Aviv 64239, Israel. E-mail: umkramer@netvision.net.il).

COMMENT. Febrile infection-related epilepsy syndrome (FIRES) or "acute encephalitis with refractory, repetitive partial seizures" (AERRPS), as preferred in Japan, is a severe epileptic encephalopathy of undefined etiology and having a poor prognosis, with cognitive impairment, refractory epilepsy, and a high mortality rate.

One case-report, a febrile acute-onset epilepsy syndrome in a girl, resembling FIRES in some respects, tested positive for missense mutation of protocadherin 19 (PCDH19) gene located at Xq22. (Specchio N et al. **Epilepsia** Nov 2011;52(11):e172-e175). The PCDH19 syndrome differs from FIRES in age at onset, ranging between 6 and 38 months and is restricted to females, whereas FIRES has an age at onset of 3-15 years and a male preponderance. The authors recommend testing for PCDH19 mutation in female acute-onset epilepsy resembling FIRES.

ADVERSE EVENTS DURING INVASIVE EEG RECORDINGS

Researchers in the Department of Paediatric Neurosurgery and Epilepsy Unit, Great Ormond Street Hospital, London, UK analyzed the hospital charts of 95 children operated on between 1995 and 2009 for medication-resistant focal epilepsy and requiring

presurgical intracranial monitoring of the EEG. Patients who underwent implantation of subdural grids and/or depth electrodes had received prior noninvasive assessment including video-EEG monitoring. Indications for invasive EEG recording included an MRI-negative focal epilepsy, discordant imaging and electroclinical data, suspected multifocal epilepsy, and seizures arising adjacent to eloquent cortex. Mean age at surgery was 10.8 years (range, 0.7-18.5 years). Mean age at the time of the first seizure was 3.4 years (range, 3 days-12 years). Mean number of seizures per month was 239.5 (range, 1-1000; SD 239). The mean number of grids or strips implanted per patient was 3 (range, 1-6). A depth electrode was placed in 31 patients.

Subdural grid recording was uneventful in 51.1% cases. Adverse events in 49% included subdural hemorrhage in 17% patients, wound infection in 14.9%, CSF leak in 10.6%, and symptomatic brain swelling in 6.4%. Adverse events were grade 1 in 19.1% (18 patients with no lengthening of hospital stay); grade 2 in 13.8% (13 with prolonged hospital stay); grade 3 in 16% (15 patients with a reduction in the Glasgow Coma Score); none was grade 4, an adverse event that would result in death. The frequency of adverse events was 20% in children <2 years of age and 50% in children >2 years of age. Of the 46 patients with adverse events, unplanned surgery had to be performed in 17 cases. No permanent neurologic deficits incurred as a result of any adverse event. Predictors of adverse events included age (brain swelling occurred in older patients >5 years); and length of recording (shorter with a complication such as hemorrhage). Functional cortical mapping with stimulation in 68 (71.6%) cases allowed identification of localized seizure onset zone in 68.9% patients. It was inconclusive in 18.9% (17 patients). The outcome of surgery was significantly related to the localizing accuracy of the invasive recording. (Blauwblomme T, Ternier J, Romero C, et al. Adverse events occurring during invasive electroencephalogram recordings in children. **Neurosurgery** December 2011;69:169-175). (Respond: Mr William Harkness MD, FRCS, Department of Paediatric Neurosurgery, Great Ormond Street Hospital for Children, Great Ormond St, London, WC1N 3JH, UK. E-mail: harknw@gosh.nhs.uk).

COMMENT. Two US neurosurgeons, Drs JG Ojemann and HL Weiner, each compliments the authors on this important contribution to the surgical management of medically refractory epilepsy. The cerebral swelling and other temporary complications of invasive monitoring are a concern. Advances in intraoperative electrocorticography in the future may lessen the need for preoperative invasive recordings.

HEADACHE DISORDERS

HEADACHE AND MOBILE PHONE USE

Researchers at Hallym University, Anyang, Korea investigated the clinical features of headache associated with mobile phone (HAMP) use among 247 medical students. Their median age was 23.6 years; 39.6% were women and 60.4% men. Following a 14-item questionnaire, individual telephone interviews were conducted with participants who reported HAMP more than 10 times during the previous year. HAMP was defined as a headache attack during MP use or within 1 hour after MP use. Of 214 (86.6%) students who completed the questionnaire, 40 (18.9%) experienced HAMP more