of normalization of the EEG at 12 months (ESX, 77%; VPA, 83%; and LTG, 64%), retention rate through the treatment period, and adverse-event rates (ESX, 25%; VPA, 29%; and LTG, 14%). Frequent causes of AED withdrawal because of adverse events were GI complaints for ESX (10%), GI complaints (5%) and alopecia (7%) for VPA, and rash for LTG (5%).

ESX, VPA and LTG are equally effective in the long-term treatment of newly diagnosed CAE patients. The onset of efficacy is faster for ESX compared with VPA or LTG. (Hwang H, Kim H, Kim SH, et al. Long-term effectiveness of ethosuximide, valproic acid, and lamotrigine in childhood absence epilepsy. Brain Dev 2012 May;34:344-348). (Respond: Dr Hee Hwang. E-mail: neuroandy@korea.com).

COMMENT. A previous double-blind, randomized, controlled clinical trial comparing the 3 drugs, ESX, VPA and LTG, found that ESX and VPA were more effective than LTG after 4-5 months of treatment. Attentional dysfunction was more common with VPA than with ethosuximide. (Glauser T et al. N Engl J Med 2010;362:790-799). The present study, extending the period of observation to 9 months, finds no significant difference in long-term effectiveness or adverse event rates of ESX, VPA and LTG. Apparently, contrary to earlier conclusions, “older (ESX) is not better!” (Vining EPG. Pediatr Neurol Briefs 2010 March;24(3):19).

YIELD OF ABNORMAL CT WITH FIRST COMPLEX FS

Physicians in Emergency Medicine, Pediatric Neurology, and Radiology at Children’s Hospital Boston, MA studied the risk of intracranial pathology requiring immediate intervention among patients presenting in the ED with a first complex febrile seizure (CFS). Of a total of 526 patients identified with a first CFS between 1995 and 2008, 268 (50.4%) had emergent head CT imaging. Four patients had a clinically significant finding: 2 had intracranial hemorrhage, 1 had ADEM, and 1 had focal cerebral edema. The risk of intracranial pathology was 4 (0.8%). Three of the 4 had other obvious findings: nystagmus, emesis, altered mental status, persistent hemiparesis, bruises suggestive of inflicted injury. Patients presenting with more than one seizure in 24 hours are at very low risk. (Kimia AA, Ben-Joseph E, Prabhu S, et al. Yield of emergent neuroimaging among children presenting with a first complex febrile seizure. Pediatr Emerg Care 2012 April;28(4):316-321). (Response and reprints: Amit A Kimia MD, Division of Emergency Medicine, Children’s Hospital Boston, 300 Longwood Ave, Boston, MA 02115. E-mail: amir.kimia@childrens.harvard.edu).

COMMENT. This study suggests that emergency neuroimaging may be unnecessary for children who present in the ED with a first CFS, uncomplicated by other acute signs of neuropathology. Focal and prolonged CFS may be more predictive of pathology than the multiple seizure type, especially when associated with prolonged postictal state.

In a retrospective study of 100 consecutive febrile seizure patient-visits to a university affiliated tertiary hospital, head CT was obtained in 18 patients at time of visit, with normal results in 17 (1 patient had mastoiditis). MRIs performed in 4 patients with CFS were normal. Of the 18 with CT scans, 4 had simple FS (5.8% of 77) and 14 had
CFS (60.9% of 23). None had neurological lesions requiring surgery. (Millichap JJ et al. Methods of investigation and management of infections causing febrile seizures. Pediatr Neurol 2008;39:381-386). CFS without neurologic signs of intracranial pathology is insufficient indication for emergent CT scan. Diagnostic criteria for CFS and indications for CT scan may require re-evaluation.

**METABOLIC DISORDERS**

**COGNITIVE OUTCOME OF INFANTS WITH POMPE DISEASE RECEIVING ENZYME-REPLACEMENT THERAPY**

Researchers at University Medical Center, Rotterdam, the Netherlands, and University of Leuven, Belgium prospectively assessed cognitive function in 10 children with classic infantile Pompe disease who had been treated with enzyme-replacement therapy (ERT) since 1999. Median age at diagnosis was 0.7 months (range 0.1-6.2 months). ERT was started at a median age of 2.3 months (range 0.1-8.3 months). Developmental scores in the first 4 years of life ranged from above average to severe delay. The type of IQ test used, severity of motor problems, speech/language delay, and age at start of ERT influenced the developmental scores. At young age poor motor functioning may interfere with reliable assessment of cognition. Scores in 5 children tested after 5 years of age ranged between normal and mild developmental delay. Nine children had hearing deficits and 7 had impaired vision. Brain imaging in 6 patients revealed periventricular white matter abnormalities in 4. (Ebbink BJ, Aarsen FK, van Gelder CM, et al. Cognitive outcome of patients with classic infantile Pompe disease receiving enzyme therapy. Neurology 2012 May 8;78:1512-1518). (Response and reprints: Prof van der Ploeg. E-mail: a.vanderploeg@erasmusmc.nl).

**COMMENT.** Pompe disease (glycogen storage disease type II; acid maltase deficiency) is an autosomal recessive, progressive metabolic myopathy due to lysosomal a-glucosidase deficiency. Enzyme activity is reduced to <1%, and glycogen stores accumulate in skeletal, cardiac, and smooth muscle, and in the brain. Pompe presents with neonatal hypotonia, macroglossia, cardiomegalia, and hepatomegalia. Patients usually die before 1 year of age with cardiorespiratory failure or aspiration pneumonia. ERT, using recombinant human a-glucosidase, improves motor development and lengthens life expectancy, but ERT does not cross the blood-brain-barrier. Glycogen is stored in the CNS and may cause cognitive deficits. This study shows that children treated with ERT who survive to school age may have normal to mildly delayed cognitive development. Testing of young children < 4 years is largely dependent on motor function. Since muscle involvement and weakness are prominent features of Pompe disease and are resistant to ERT, cognitive development will be underestimated in children younger than 5 years.