

developed status epilepticus. An EEG was compatible with diffuse encephalopathy or encephalitis. Treatment with acyclovir was without benefit. MRI revealed cerebellitis with swelling and multiple enhancing lesions. Severe increased intracranial pressure with tonsillar herniation and hydrocephalus responded to dexamethasone. Hemophagocytic lymphohistiocytosis (HLH) was suspected, and treatment with dexamethasone, etoposide, and cyclosporin A (HLH-94 protocol) resulted in a dramatic clinical response, with normal CSF and EEG, and improvement in the MRI. Cord blood stem cell transplantation was performed 10 months later, with initial infectious complications and neurologic sequelae (seizures, behavior disorder, and visual field defects), but subsequent clinical improvement. Genetic studies with DNA sequencing showing a mutation in the perforin gene confirmed the diagnosis of familial HLH. (Astigarraga I, Prats JM, Navajas A et al. Near fatal cerebellar swelling in familial hemophagocytic lymphohistiocytosis. *Pediatr Neurol* 2004;30:361-364). (Respond: Dr Astigarraga, Pediatric Oncology Unit, Department of Pediatrics, Hospital de Cruces, 48903 Barakaldo, Bizkaia, Spain).

COMMENT. Familial histophagocytic lymphohistiocytosis (HLH) is often triggered by infection, and varicella preceded the onset of neurologic symptoms in this case. The disease is characterized by activation of T cells and macrophages leading to an inadequate immune response. CNS involvement is common and symptoms include bulging fontanel, neck stiffness, seizures, ataxia, visual disturbance, hemiplegia, and coma with increased intracranial pressure. In the above report, cerebellar signs preceded the systemic presentation with hepatosplenomegaly and cytopenia. HLH should be suspected in children with progressive encephalopathy and pancytopenia of unknown cause, and therapy instituted early. Dexamethasone, etoposide, and cyclosporine followed by stem cell transplantation are effective, and result in a 3-year probability survival in 62% cases.

HLH or ‘hemophagocytic syndrome’ with lamotrigine. (Yang Y-C, Jou S-T, Chang Y-H et al. *Pediatr Neurol* 2004;30:358-360). An 8 year-old male with porencephaly and epilepsy unresponsive to topiramate and valproate developed a skin rash 2 weeks after beginning antiepileptic treatment with lamotrigine. One month later, impaired liver function, pancytopenia and hemophagocytosis without evidence of infection led to a diagnosis of HLH. The blood count and liver function improved dramatically when AEDs were discontinued and IV immunoglobulin and steroid were administered. Pancytopenia during treatment with new AEDs such as lamotrigine should alert to a possible HLH. Phenytoin may also trigger HLH, and valproate, also suspect in this case, may elevate blood levels of lamotrigine.

TRAUMATIC DISORDERS

RISKS OF BRAIN INJURY AFTER BLUNT HEAD TRAUMA

The association of loss of consciousness (LOC) and/or amnesia with traumatic brain injury (TBI) identified on CT and TBI requiring acute intervention was evaluated in 2043 children <18 years old enrolled prospectively in a level 1 trauma center ED at University of California, Davis School of Medicine, CA. A documented history of LOC and/or amnesia was obtained in 801 (39%). Of 745 with LOC and/or amnesia and CT, 70 (9.4%) had TBI

identified on CT, whereas only 11 (2.7%) of 414 without LOC and/or amnesia had CT evidence of TBI; 9.6% of patients with LOC had TBI requiring acute intervention compared to 1% of those without LOC. Of 142 patients with isolated LOC without other signs or symptoms of TBI, none had CT evidence of TBI and none required acute intervention. Isolated LOC and/or amnesia, without other findings suggestive of TBI, are not predictive of TBI on CT or TBI that requires acute intervention and should eliminate the need for CT. Isolated LOC is defined by the absence of vomiting, seizure, headache, skull fracture, altered mental status, neurologic deficit, or scalp hematomas. (Palchak MJ, Holmes JF, Vance CW et al. Does an isolated history of loss of consciousness or amnesia predict brain injuries in children after blunt head trauma? **Pediatrics** June 2004;113:e507-e513). (Reprints: MJ Palchak MD, Division of Emergency Medicine, University of California, Davis Medical Center, 2315 Stockton Blvd, Sacramento, CA 95817).

COMMENT. The diagnostic value of CT in the evaluation of a child with blunt head trauma must be weighed against the disadvantage of the transport of the patient, radiation exposure, possible need for sedation, and costs. Nevertheless, several guidelines recommend CT for all children with a history of LOC after blunt head trauma. This study minimizes the value of CT in cases of LOC with blunt head trauma, especially in patients without other signs or symptoms of head trauma. The authors caution that the findings in their center may not be generalized to all centers, and the data are insufficient for a meaningful analysis of cases less than 2 years of age and those secondary to child abuse. External validation of the results is suggested.

Mild head injury may result in cognitive deficits and behavior disorders, and a normal CT after head injury is predictive of a good prognosis and lack of subsequent mental deterioration. (Davis RL et al. **Pediatrics** 1995;345-349; **Ped Neur Briefs** April 1995).

METABOLIC DISORDERS

BRAIN DAMAGE IN GLYCOGEN STORAGE DISEASE TYPE I

The occurrence of brain damage in 19 patients (13 girls and 6 boys) with glycogen storage disease type I (GSDI) was evaluated at the Università "Federico II", Naples, Italy. Performance ability as measured on Wechsler IQ tests showed lower scores in patients compared to controls ($p<0.05$). The prevalence of abnormal EEG findings (26.3% vs 2.6%), VEPs (38.4% vs 7.7%), SEPs (23% vs 0%), and BAEPs abnormalities (15.7% vs 0%) was higher in patients than controls ($p<0.05$). MRI showed abnormalities (dilated occipital horns, hyperintensity of subcortical white matter) in 8/14 (57.1%) patients and none of controls. Performance ability and BAEP abnormalities correlated significantly with the frequency of admissions for hypoglycemia. EEG abnormalities correlated with poor dietary compliance. (Melis D, Parenti G, Casa RD et al. Brain damage in glycogen storage disease type I. **J Pediatr** May 2004;14:637-642). (Reprints: Professor Generoso Andria, Dipartimento di Pediatria, Università "Federico II," Via Sergio Pansini 5, 80131 Naples, Italy).

COMMENT. GSDI is characterized by hypoglycemia, hyperlactic acidemia, and hepatorenal enlargement. Neonatal and recurrent hypoglycemia play a key role in causing brain damage in GSDI, and dietary compliance is essential in treatment.