hypertension in younger patients may have a different mechanism, no female sex preponderance, and no increased frequency of obesity, when compared to adolescent patients.

METABOLIC DISORDERS

INFANTILE NEUROGLYCOPENIA

Participant 1, a 23-year-old woman with congenital hypoglycemia (hyperinsulinism); participant 2, a 16-year-old boy with genetic mutation of the cerebral glucose transporter type 1 (GLUT1 deficient); and participant 3, the 23-year-old healthy twin sister of participant 1 as a control, received a neurologic examination, PET scan, and neuropsychological evaluation, in a study at Neurological Institute, New York, NY. Pt 1 with a residual encephalopathy at 23 years showed hypertonicity, ataxia, dysarthria, hyperreflexia with ankle clonus and Babinski signs. Pt 2 had residual epileptic encephalopathy at 16 years, he received special education, and the neurologic examination was identical to that of pt 1. Treatment with diazoxide increased glycemia and resulted in improved alertness and psychomotor coordination. Pt 3 had a normal neurologic exam and PET scan. PET scans of Pts 1 and 2 were identical, showing hypometabolic activity in parietal, temporal, and thalamic regions, and enhancement of uptake in the basal ganglia, results typical of GLUT1 deficiency. Wechsler scale IQ scores were Pt 1, 64; Pt 2, 49; and Pt 3, 104. Pts 1 and 2 had impaired cognitive skills compared to the healthy twin, Pt 3.

The syndrome of neuroglycopenia is characterized by infantile epilepsy, mental retardation, abnormal neurologic exam and PET scan, all caused by a persistent decrease in glucose supply to the developing brain. The glycopenia may be the result of congenital hypoglycemia or GLUT1 deficiency, and both are treatable diseases. Patients with a milder GLUT1 phenotype are responsible for the familial autosomal dominant transmission of the disease. Diagnosis is made by the typical neurologic manifestations and lumbar puncture showing hypoglycorrhachia. (Pascual JM, Wang D, Hinton V et al. Brain glucose supply and the syndrome of infantile neuroglycopenia. **Arch Neurol** April 2007;64:507-513). (Respond: Juan M Pascual MD, PhD, Department of Neurology, University of Texas Southwestern Medical Center, 5323 Harry Hines Blvd, Mail Code 8813, Dallas, TX 75390).

COMMENT. Neuroglycopenia may be caused by GLUT1 deficiency or infantile hypoglycemia. The neurologic, psychological, and functional imaging characteristics are similar for both disorders. Early hyperexcitability and seizures are associated with encephalopathy and pyramidal and cerebellar dysfunction. Glucose may have a dual function: 1) as a fuel (energy supply), hypoglycemia resulting in acute neurologic deficits, and 2) as a necessary substrate for normal thalamocortical maturation and development of the nervous system.