

intracranial hemorrhage and ischemic infarctions have occurred among children with immune thrombocytopenia or arteritis. Acute hemiplegia secondary to a large infarct is described in a 16-month-old infant with congenitally acquired HIV infection (Kugler SL et al. Ped. Neurol May/June 1991; 7:207-10). School achievement and tasks requiring motor speed, attention and concentration were impaired at 4-8 years of age in 15 HIV-1 seropositive children infected through neonatal blood transfusion (Cohen SE et al. Pediatrics July 1991; 88:58-68).

CT scans may demonstrate enlarged CSF spaces and bilateral symmetrical calcification of the basal ganglia and frontal white matter 17-61% (Belman A L et al. A J D C 1988; 142:29-35). Wiley C A et al. report neocortical damage during HIV infection in an autopsy study of adults (Ann Neurol June 1991; 29:651-657). Wzdicks E F M et al. report a fatal disseminated hemorrhagic toxoplasmotic encephalopathy as the initial manifestation of AIDS in a 28 year-old woman (Ann Neurol June 1991; 29:683-686).

#### **MYOPATHY IN ZIDOVUDINE TREATED HIV INFECTION**

Elevations in serum CK levels in 80% of 87 children enrolled in a multi-centered trial of oral zidovudine treatment for symptomatic HIV infection are reported from the Department of Pediatrics, University Medical Center, Durham, NC. A myopathy developed in a 3 year old girl with acquired HIV infection who received long-term zidovudine therapy. Muscle biopsy was consistent with a non-inflammatory myopathy but a beneficial response to prednisone suggested an inflammatory component. Severe neurologic impairment related to HIV encephalopathy masked the early signs of proximal muscle weakness. Electron microscopy revealed no mitochondrial changes. She had received zidovudine (180 mg/m<sup>2</sup> every 6 hours) for two years. Therapy was discontinued because of fever and neutropenia. The CK level fell from a peak of 25,945 IU/L to 185 IU/L at the time of discharge. (Walter E B et al. Myopathy in human immunodeficiency virus-infected children receiving long-term zidovudine therapy. J Pediatr July 1991; 119: 152-155).

**COMMENT.** Myopathy associated with HIV infection is usually an inflammatory polymyositis with proximal muscle weakness and an elevated CK level. It may also be non-inflammatory or linked to treatment with antiretroviral agents such as zidovudine. Myopathy associated with zidovudine is characterized by reversible changes in muscle mitochondria, whereas mitochondrial features are normal in specimens from HIV infected patients not receiving zidovudine.

Low-dose zidovudine in 65 children with HIV-1 infection acquired in the perinatal period was well tolerated in a study reported from the Pediatric Neurology Division, Bicetre Hospital, Institut National de la Sante et de la Recherche Medicale, France (Blanche S et al. Pediatrics Aug 1991; 88:364-370). The dosage (400 mg/m<sup>2</sup> per day)

was lower than that used in the above myopathy case report (720 mg/m<sup>2</sup> daily). Moderate elevations in CK levels are to be expected with zidovudine therapy, and muscle biopsy and EMG may be reserved for children with clinical myopathy or markedly elevated CK levels.

### **GUILLAIN-BARRE SYNDROME: AGE VARIATIONS**

The medical records of 83 children with Guillain-Barre Syndrome were reviewed with reference to the age associated changes at the Department of Pediatrics, University of Tokyo, Japan. Children between 3 and 9 years were most susceptible and summer was the most common time of onset. Limb pain was the initial symptom for 53% of all patients and for 24% in the younger age group ( $P < 0.05$ ). Muscle weakness was the most frequent initial symptom in the younger age group. Upper respiratory infections were the most common preceding illnesses; the interval between the previous illness and the onset of disease was significantly shorter for the older age group. The total duration of the illness was shorter in older children. Cranial nerve involvement occurred in 1/3 of children under 5 years of age and in the same proportion of children over 5 years. In the younger age group bulbar nerves were most commonly affected whereas the facial nerve was involved most frequently in the older age group. Patients with cranial nerve involvement suffered more frequently from respiratory insufficiency; the ages of those affected were distributed evenly between the two groups. The differences in initial symptoms and in interval between preceding illnesses and onset of the disease seemed to be related to the maturational processes of peripheral nerve and spinal root ganglia. It is speculated that the stage of the myelinogenic process might be important in the explanation of age associated alterations in symptomatology (Sakakihara Y, Kamoshita S. Age associated changes in the symptomatology of Guillain-Barre syndrome in children. Dev Med Child Neurol July 1991; 33:611-616).

**COMMENT.** The incidence of Guillain-Barre Syndrome increases with age but 25% of cases occur in childhood and adolescence. Since nerve conduction velocity and the thickness of the myelin sheath in peripheral nerves reaches adult levels at around five years of age, some variation in symptomatology between young and older children might be expected. Electrodiagnostic tests have been shown to corroborate the neuropathologic sequence in the Guillain-Barre Syndrome and these findings may be modified by age and maturation (Ropper AH et al. Arch Neurol 1990; 47:367; McFarland HR Arch Neurol July 1991; 48:678)

## **DEVELOPMENTAL DISORDERS**

### **LISSENCEPHALY: CLINICAL AND MRI FINDINGS**

Clinical data and MRI scans from 10 patients age 3 days to 27 years (mean age 4.6 years) with lissencephaly were reviewed in the Departments of Radiology, Neurology and Pediatrics, University of California, San Francisco,