COMMENT. In infants and young children with brain tumors and few signs or subjective complaints, the knee-chest posture might be helpful in the clinical suspicion and diagnosis of increased intracranial pressure. This posture is also assumed by children with cyanotic heart disease and is a characteristic sign of acrodynia.

DEGENERATIVE DISEASES

FOCAL TUMOR-LIKE DEMYELINATING LESIONS

Thirty-one patients with large, focal cerebral demyelinating lesions are reported from the University of Kansas School of Medicine, Kansas City. The lesions presented clinically and radiologically as brain tumors or as multiple cysts. The demyelinating pathology was established through biopsy and a significant improvement with corticosteroid therapy. Four patients were children, ages 8 - 12 years, and one was an adolescent, aged 18. One young adult had received an influenza vaccine 10 days before the onset of symptoms. The clinical course favored postinfectious/postvaccination encephalitis, but the tumor-sized masses of demyelination were atypical. (Kepes JJ. Large focal tumor-like demyelinating lesions of the brain: intermediate entity between multiple sclerosis and acute disseminated encephalomyelitis? A study of 31 patients. Ann Neurol Jan 1993; 33: 18-27). (Correspondence: Dr Kepes, Department of Pathology and Oncology, University of Kansas College of Health Sciences and Hospital, 39th and Rainbow Blvd, Kansas City, KS 66103).

COMMENT. The author rules out Schilder's disease, a progressive childhood diffuse sclerosis, which often presents with asymmetrical, large focal hemispheric lesions and smaller lesions, sometimes difficult to differentiate from multiple sclerosis. (See Greenfield's Neuropathology, Baltimore, Williams and Wilkins, 1963). Many reported cases of Schilder's disease are now thought to represent either adrenoleukodystrophy or acute forms of multiple sclerosis (Menkes JH. Textbook of Child Neurology. 3rd ed. Philadelphia, Lea & Febiger, 1985).

MRI IN INFANTILE NEUROAXONAL DYSTROPHY

The MRI findings in four children aged 3 to 10 years with infantile neuroaxonal dystrophy are reported from the Chiba Children's Hospital, Japan. T_2 - weighted images showed bilateral diffuse hyperintensity of the cerebellar cortex and cerebellar atrophy. Autopsy changes in one patient included cerebellar atrophy, ventricular dilatation, spheroids in the gray matter, loss of neurons, axonal swellings, and extensive astrogliosis. (Tanabe Y et al. The use of magnetic resonance imaging in diagnosing infantile neuroaxonal dystrophy. Neurology Jan 1993; 43: 110-113). (Reprints: Dr Y Tanabe, Division of Neurology, Chiba Children's Hospital, 579-1 Heta-cho, Midori-ku, Chiba 266, Japan).

COMMENT. The authors comment that these MRI findings may be specific to infantile neuoaxonal dystrophy and permit early diagnosis. The clinical picture includes upper and lower motor neuron deficits, including progressive weakness, difficulty in walking, hypotonia, muscle atrophy, hyperactive reflexes, Babinski signs, and optic atrophy. Diagnosis is confirmed by peripheral nerve biopsy, which shows globular swellings on the axons. Axonal swelling may also occur in Hallervorden-Spatz disease, Friedreich's ataxia, and other neurodegenerative diseases, however.

MRI IN INFANTILE KRABBE DISEASE

Serial MRI findings paralleling the clinical deterioration of a year-old child with Krabbe disease are reported from the University of Rochester Medical Center, New York. At 13 months, an abnormal high signal on T_2-weighted images in the frontoparietal white matter and cerebellar white matter was correlated with severe developmental delay, increased tone, and hyperextension (stage 2). At 32 months, MRI changes had progressed to include central and cortical atrophy, decreased white matter volume, thalamic and caudate atrophy, and abnormal high signal in all motor tracts except the anterior limb of the internal capsule. Clinically, the child was in stage 3, with spasticity of the upper extremities, peripheral neuropathy, and no response to auditory or visual stimuli. (Farley TJ et al. Serial MRI and CT findings in infantile Krabbe disease. Pediatr Neurol Nov/Dec 1992; 8: 455-458). (Correspondence: Dr Ketonen, Dept Radiology, University of Rochester Medical Center, Box 648, 601 Elmwood Ave, Rochester, NY 14642).

COMMENT. The 3-stages of Krabbe disease described by Hagberg begin with hyperirritability, restlessness, and frequent crying. Convulsions may develop, induced by sensory stimuli. Development becomes delayed, tone is increased, and later, reflexes are difficult to elicit and are absent in the lower limbs. Terminally, the infant is flaccid and blind. CSF protein is elevated and nerve conduction is delayed. The absence of the enzyme galactosyl ceramide B-galactosidase in leukocytes and skin fibroblasts is diagnostic, a test result common to all variants and stages of Krabbe disease, including antenatal.

BIOCHEMICAL MARKER FOR MENKES DISEASE

Plasma and CSF levels of catechols in 10 patients with Menkes disease, ranging in age from 9 days to 27 months, were compared with control groups and patients with congenital absence of dopamine-B-hydroxylase (DBH) at the National Institutes of Health, Bethesda, MD. The neurochemical pattern in Menkes disease patients was characterized by high dihydroxyphenylalanine (DOPA), dopamine (DA), and dihydroxyphenylacetic acid (DOPAC) levels,