MOYAMOYA DISEASE

Sixteen children with moyamoya disease and involvement of the posterior cerebral artery are reported from the Departments of Neurophysiology, Neurosurgery, and Radiology, Faculty of Medicine, Kyushu University, Fukuoka, Japan. Eight patients had complete occlusion of posterior cerebral arteries and the other eight had nonocclusive disease. All patients showed patent ophthalmic arteries bilaterally. Pattern-reversal visual-evoked potentials showed abnormalities in 75% of the posterior cerebral artery occlusive group and no abnormalities in the nonocclusive group. Abnormalities were also found in positron emission tomography, computed tomography, and the clinical examination of the visual fields. The authors concluded that the pattern-reversal visual-evoked potentials was the most practical means to explore posterior cerebral artery occlusion in the course of moyamoya disease. (Tashima-Kurita S et al. Moyamoya disease. Posterior cerebral artery occlusion and pattern-reversal visual-evoked potential. Arch Neurol May 1989; 46:550-553).

COMMENT. Moyamoya disease is characterized by progressive occlusion of cerebral arteries and predominantly the anterior and middle cerebral arteries. The incidence of posterior cerebral artery involvement and visual disturbances is approximately 25% and the clinical manifestations include decreased visual acuity, homonymous hemianopsia, constriction of the visual fields, and scintillating scotoma.

METABOLIC DISORDERS

CNS COMPLICATIONS OF CYSTINOSIS

Fourteen patients with cystinosis, eight males and six females ranging in age from 13 to 24 years (mean 18.1 years) were examined for neurological involvement at the National Institute of Neurologic and Communicative Disorders and Stroke, the National Institutes of Health Clinical Center, Bethesda, Maryland. Two patients had neurological symptoms, including bradykinesia, dementia and spasticity, and behavioral and cognitive disturbances; 12 patients had CT evidence of generalized cerebral atrophy; two had multifocal intracerebral mineralization on CT scan; two had abnormal electroencephalograms and only one patient was entirely normal. Patients with neurologic symptoms or markedly abnormal CT scans were older and had a longer interval between their initial renal transplantation and the examination at follow-up than those patients who were normal or who had only mild cerebral atrophy. The neurologic and neuropsychometric abnormalities correlated with the degree of roentgenographic abnormality. The patients with nervous system abnormalities were not distinguished by patterns of medication use or the relative severity of cystinosis. The differential diagnosis included other complications from renal failure, dialysis and immunosuppression. (Fink JK et al. Neurologic complications in long-standing nephropathic cystinosis. Arch Neurol May 1989; 46:543-548).