

and congenital porencephaly should be evaluated with MRI for coexistent mesial temporal sclerosis. Hippocampal formation atrophy is the more likely origin for the seizures in patients with dual pathologies, particularly when the EEG shows localization to the temporal lobe.

Quantitative MRI of the hippocampus (Van Paesschen W et al. Ann Neurol Nov 1997;42:756-766), and **Proton magnetic resonance spectroscopic imaging** (Cendes F et al. Ann Neurol Nov 1997;42:737-746) were used in the presurgical evaluation of temporal lobe epilepsy at the National Hospital, London, UK, and Montreal Neurological Institute, Canada.

CEREBELLAR DISORDERS

CEREBELLAR STRUCTURAL ABNORMALITIES AND GENETICS

The etiology and incidence of known metabolic and hereditary disorders associated with unilateral or bilateral structural cerebellar abnormalities, defined by CT and/or MRI, were determined in 78 children examined at the University Hospital Aachen, Germany, and Katholieke Universiteit Leuven, Belgium. Lesions were bilateral in 62 and unilateral in 16, both cerebellar hemispheres were involved in 38, the vermis in 15, and pontocerebellar in 9. Hemisphere atrophy was static in 10 and progressive in 28. MRI was superior to CT in definition of lesions. Genetic/metabolic causes were found in more than half the cases of ponto-cerebellar hypoplasia or progressive cerebellar atrophy, but in none with unilateral cerebellar lesions. These included amino and organic acidurias, lactic acidosis, lysosomal and peroxisomal disorders, Menkes kinky hair disease, molybdenum cofactor deficiency, and autosomal dominant ataxias. Other causes of cerebellar and pontocerebellar hypoplasia included intrauterine ionizing radiation, phenytoin exposure, cytomegalovirus, chromosomal syndromes, hypogonadism, Ito's hypomelanosis, and carbohydrate-deficient-glycoproteins syndromes. Investigations should include EEG, EMG and NCS, abdominal ultrasound, urine screening for amino and organic acids, and blood tests for acanthocytes, liver function, protein electrophoresis, ammonia, lactate and pyruvate, copper and ceruloplasmin, immunoglobulins, VLC fatty acids, and glycoproteins. An overview of the literature is also presented. (Ramaekers VTh, Heimann G, Reul J, Thron A, Jaeken J. Genetic disorders and cerebellar structural abnormalities in childhood. Brain Oct 1997;120:1739-1751). (Respond: Dr V Th Ramaekers MD, Department of Paediatrics, Medizinische Einrichtungen der RWTH, Pauwelsstrasse 30, D-52057 Aachen, Germany).

COMMENT. Pontocerebellar hypoplasia or progressive cerebellar atrophy defined by MRI is an indication for biochemical and neurophysiological tests for hereditary or degenerative neurological disorders.

Focal cerebellar lesions and associated learning impairments were detected in 8 patients cf 6 control subjects tested in a serial reaction-time task at the Catholic University, and University of Rome 'La Sapienza', Rome, Italy. (Molinari M et al. Brain Oct 1997;120:1753-1762). Cerebellar patients had longer reaction times than controls when stimuli were presented in sequence.

DEMYELINATING DISORDERS

OPTIC NEURITIS AND RISK OF MULTIPLE SCLEROSIS

Risk factors for the development of multiple sclerosis (MS) were