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 neurophsiological 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

determined by follow-up (mean 22 yrs) of 79 children who presented with a diagnosis of optic neuritis (ON) at the Mayo Clinic between 1950 and 1988. The incidence of MS was 13% by 10 yrs follow-up and 19% by 20 yrs. Of the patients who developed MS, 7 of 15 (47%) had symptoms within the first year after recovery from ON; one developed MS after 31 years interval. Four had clinical signs of Devic's disease. Bilateral sequential or recurrent ON increased the risk of MS, whereas unilateral ON was associated with a low risk of MS. Of 37 children with unilateral ON, only 2 (5%) developed MS. The presence of recent infection within 2 weeks of onset of ON decreased the risk of MS. Childhood onset ON had a lower risk of progression to MS compared to ON in adults. (Lucchinetti CF, Kiers L, O'Duffy A, et al. Risk factors for developing multiple sclerosis after childhood optic neuritis. Neurology Nov 1997;49:1413-1418). (Reprints: Dr M Rodriguez, Department of Neurology, 200 First Street SW, Rochester, MN 55905).

COMMENT. The risk of developing multiple sclerosis after optic neuritis in childhood is lower than in adult-onset ON and increases with length of follow-up from 13% after 10 years to 22% by 30 years. Bilateral ON carries a greater risk of MS, whereas infection preceding the ON lessens the risk of MS.

In a previous report from Gottingen, Germany (Hanefeld FA, 1995), onset or relapse of MS was preceded by a nonspecific infection, usually an URI, in >50% cases. Of 8 presenting with ON, 4 developed MS within 2 years. (See Progress in Pediatric Neurology III, PNB Publ, 1997;551-554, for further articles on childhood MS).

CHRONIC INFLAMMATORY POLYRADICULONEUROPATHY

The long-term clinical course of 12 children with idiopathic chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is reported from the University of Michigan, Ann Arbor, and Pennsylvania State University/Hershey Medical Center, Hershey, PA. Data on 62 adults with CIDP previously reported from these centers were used for comparison. In children aged 3 to 17 years, boys and girls were equally affected. Illness duration was 1 to 25 years. In contrast to adults with CIDP, children were more likely to relapse but none was progressive; 83% childhood cases relapsed cf to 35% adults. All were treated with prednisone, plasma exchange, or IV immunoglobulin, and recovery from each relapse was excellent. Only 2 with precipitous onset required ventilatory support. Adults showed a more variable outcome. (Simmons Z, Wald JJ, Albers JW. Chronic inflammatory demyelinating polyradiculoneuropathy in children: II. Long-term follow-up, with comparison to adults. Muscle & Nerve December 1997;20:1569-1575). (Respond: Dr Zachary Simmons, Division of Neurology, Pennsylvania State University College of Medicine, Hershey Medical Center, Hershey, PA 17033).

COMMENT. According to the above study, the long-term prognosis for children with CIDP is excellent, but relapses are to be expected. Those with a slowly progressive onset have a similar outcome to the acute onset cases, and none develops a severe disability. A gradual weaning from therapy is often successful. These findings are in contrast to the experience at Washington University, St Louis, MO, and the Royal Children's Hospital, Melbourne, Australia, as reviewed in Ped Neur Briefs Aug 1996 (Progress in Pediatric Neurology III, 1997;pp360-362). Nevo, Pestronk et al reported that childhood onset CIDP has in general a poor prognosis, the majority relapsing and having residual weakness, and attempts to withdraw steroids frequently unsuccessful.