The ages ranged from 8 mos to 58 yrs with a childhood preponderance and 28% below 6 years. The male:female ratio was 2.65:1. Only 7% had a history of febrile convulsions. Complex partial seizures were the most frequent manifestation of HWE (67%) and generalized tonic-clonic seizures occurred in 33%. Spontaneous non-reflex epilepsy followed or preceded the onset of HWE in 30%. A positive family history of epilepsy was obtained in 22% and for HWE in only 7%. The avoidance of the hot water stimulus should be supplemented with anticonvulsant medication in therapy. (Satishchandra P et al. Hot-water epilepsy: a variant of reflex epilepsy in Southern India. Epilepsia Jan/Feb 1988;29:52-6).

**COMMENT.** The mechanism of HWE is unclear. A hot-air stimulus to the heads of patients failed to induce attacks. A kindling effect has been induced in rats by repeated exposure of the head to hot water (Klanenberg BJ, Sparber SB. Epilepsia 1984;25:292). Hot water applied to the abdomen induces fever and changes in cortical electrical activity of cats and kittens (Kashiwase Y. Brain Nerve (Tokyo) 1962;14:698). The body temperature of patients in the present study is not documented and fever induced by the hot water stimulus may explain some cases, especially in younger children.

Absence epilepsy evoked by thinking or talking about driving an automobile is an unusual example of reflex epilepsy also reported in the current issue of Epilepsia (Bencze KS et al. of the Dept of Neurology, University of South Florida, Tampa, FL).

### DEGENERATIVE AND METABOLIC DISORDERS

#### MONOAMINE METABOLITES IN RETT SYNDROME

Cerebral metabolites of noradrenaline, dopamine and serotonin, y-aminobutyric acid, and 23 amino acids were present in normal concentrations in the CSF of 5 girls with Rett syndrome studied in the Depts of Pharmacology and Therapeutics, and Dept of Paediatrics, University of British Columbia, Vancouver, Canada. The authors doubt that any biochemical abnormalities have been clearly established as characteristic of the syndrome. (Perry TL, Dunn HG et al. Cerebrospinal fluid values for monoamine metabolites, y-aminobutyric acid and other amino compounds in Rett syndrome. J Pediatr Feb 1988;112:234-8).

**COMMENT.** A previous report of low CSF levels of monoamine metabolites in patients with Rett syndrome (Zoghbi HY et al. N Engl J Med 1985;313:921) is not supported by the present study. Hyperammonemia reported originally by Rett is another suggested biochemical basis for the syndrome unconfirmed in subsequent reports. The lack of uniformity of these findings suggests that Rett syndrome is a nonspecific entity with more than one etiology.

#### INFANTILE REFSUM DISEASE

Two patients with infantile Refsum (phytanic acid storage) disease were treated at 9 mos and 5½ years of age with a low phytanic acid diet and the effects studied over a 2-yr period or longer in the Depts of Chemical Pathology, Neurology and Histopathology, Adelaide Children's Hospital, and the Dept of Neurology, Prince of Wales Children's Hospital, New South Wales,
Australia. Elevated phytanic and pipecolic acid plasma levels were significantly decreased but very long chain fatty acids remained grossly abnormal after 6 wks or longer on the diet consisting of skimmed milk powder, white meats, fish, root vegetables, some fruits and supplementary vitamins. Growth, motor skills, self-mutilating behavior, intention tremor and nystagmus improved, although ataxia and hypotonia persisted in the older child who developed peripheral neuropathy with slowed motor and sensory nerve conduction velocities. Impaired visual acuity with optic atrophy and retinitis pigmentosa and sensorineural deafness persisted. Head growth continued at the original 25th and 3rd low percentiles. Electronmicroscopy of liver biopsies before and after dietary treatment showed an increase in inclusion bodies, and peroxisomes and lysosomes were present. (Robertson EF et al. Treatment of infantile phytic acid storage disease: clinical, biochemical and ultrastructural findings in two children treated for 2 years. Br J Pediatr Feb 1988;147:133-42).

**COMMENT.** Infantile Refsum syndrome is characterized by retinitis pigmentosa, sensorineural deafness, developmental delay, hepatomegaly and dysmorphic features. The older of the 2 present cases showed chronic polyneuropathy, ataxia and intention tremor, typical of the classical form of Refsum disease-heredopathia atactica polyniuritiformis. The exclusion of phytanic acid from the diet of affected adults has been successful in lowering plasma phytanic acid levels, improving peripheral nerve function and arresting the progress of visual and hearing defects.

**LEARNING DISORDERS**

**ZINC DEFICIENCY AND DYSLEXIA**

An association between dyslexia and low concentrations of zinc in sweat has been demonstrated in a study of 26 children aged 6-14 yrs recruited from those attending the Dyslexia Institute, Staines, Middlesex, and the Hornsby Learning Centre, London. They were paired with their non-dyslexic school friends, who were matched for age and sex and had no obvious allergies, illnesses or behavior disorders. Sweat from the skin of the back and hair from the occipitonoachal region were collected for analyses of trace minerals at the Biolab Medical Unit, London. W1N3PF (correspondence to Dr. Davies).

Zinc concentrations in sweat of dyslexic children (5.4 umol/l) was 66% that in controls (8.0 umol/l, P=.0001). Higher concentrations of copper, lead and cadmium and no differences in zinc concentrations were found in hair of dyslexics compared with controls. (Grant ECG et al. Zinc deficiency in children with dyslexia: concentrations of zinc and other minerals in sweat and hair. Br Med J 27 Feb 1988;296:607-9).

**COMMENT.** Zinc in sweat may be a more useful guide to clinical zinc deficiency than hair or serum concentrations (Davies S. Sci Total Environ 1985;42:45 – quoted above). The authors propose that zinc deficiency in parents may possibly predispose to familial dyslexia. Zinc deficiency can be due to nutritional factors, inherited defects in zinc metabolism, and several disease states (Nutrition, Diet and Your Child's Behavior. C C Thomas, Springfield, 1986). The high phytate content of protein and the fiber in certain cereals decrease the availability of zinc in persons who eat primarily cereals and little meat. Alcoholism, malabsorption, kidney disease, and sickle-cell anemia predispose to zinc deficiency. The reported possible association of maternal zinc-deficient diets with