

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 phytanic acid storage disease: clinical, biochemical and ultrastructural findings in two children treated for 2 years. Eur 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 polyneuritiformis. The exclusion of pytanic 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 DYSLLEXIA

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 occipitonalchal region were collected for analyses of trace minerals at the Biolab Medical Unit, London W1N3FF (correspondence to Dr. Davies).

Zinc concentrations in sweat of dyslexic children (5.4 umol/l) was 66% that in controls (8.0 umol/l, $P=0.001$). 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

developmental defects in fetal brain is of interest in relation to the brain malformations recorded on CT scans of occasional children with dyslexia (Galaburda AM et al. Ann Neurol 1985;18:222).

Hair zinc levels of urban toddlers were lower than rural toddlers, especially in summer and in those with frequent upper respiratory tract infections, in a study from North Rhine-Westphalia, Fed Rep Germany (Lombeck I et al. Eur J Pediatr Feb 1988;147:179). Environmental and seasonal factors and age, sex and infection affect the variability in zinc hair content in addition to dietary factors. Caution in the interpretation of hair analyses is stressed.

MOVEMENT DISORDERS

TOURETTE'S SYNDROME

The prevalence of Gilles de la Tourette's syndrome (TS) in school children from Monroe County, New York, was examined in the Depts of Psychiatry, Neurology and Pediatrics, University of Rochester Medical Center, Rochester, NY. Forty-one TS patients were detected with an estimated prevalence of 28.7 per 100,000. Twenty (50%) of the 41 children had obsessive ideas or ritualistic motor behaviors associated with obsessive ideas. These included rituals to make sure the body was symmetrical or balanced, mental arithmetic games, touching of objects to ward off bad luck, or repetitively placing objects. Only 3 had a diagnosable obsessive-compulsive disorder. Eleven children had attention deficit disorder with hyperactivity (ADHD); of 10 who had received methylphenidate, one developed tics after 18 months of therapy and movements almost completely ceased when drug was discontinued. Eleven had insomnia and seven had self-harming behaviors, including lip biting, sticking pins in the skin, and burning fingers on hot objects. Twenty patients had complex vocalizations including coprolalia, echolalia and stuttering. On neurologic examination, 12 showed subtle abnormalities or soft signs including synkinesis, impaired rapid alternating movements, and inability to hop, and one had significant postural and motor defects associated with microcephaly and growth retardation. Thirty-seven were male and four were female. Fifty-six percent had a positive family history of TS or tics. TS was a mild disorder requiring no drug treatment in 23 (56%) patients. Thirteen of those who received haloperidol (0.5-2.5mg daily) were benefited and 5 patients were uncontrolled. (Caine ED et al. Tourette's syndrome in Monroe County school children. Neurology March 1988;88:472-5).

COMMENT. Tourette's syndrome is manifested by the onset of recurrent multiple motor tics and involuntary vocal tics in childhood. The incidence of reported cases was low until the 1960s and the condition was generally omitted from the index of textbooks of neurology. Increased public awareness of the disorder and recognition of organic in addition to functional psychiatric causes have led to an increase in apparent prevalence. Formerly, the combination of tics, coprolalia and barking were required for diagnosis whereas recently, patients with simple habit spasms are sometimes included in collected series. The exact etiology is unknown although organic factors are suspected. About 10% have a history of previous head injury or neonatal asphyxia (Erenberg G et al. Cleve Clin Q 1986;53:127). Many have learning problems and behavior disorders for which psychostimulant medication may be indicated. Methylphenidate should be withheld or used with caution at lower dose levels in children with tics or a family history of Tourette's syndrome.