West Virginia University, (Neurologic Clinics August 1990, W. B. Saunders, Philadelphia) recommend 150 U/m²/day for a total of 6-8 weeks and the same dosage on alternate days for a further 6-8 weeks followed by tapering for a total treatment period of 4-6 months. These larger doses are associated with an increased frequency of serious side effects from ACTH. My own preference has favored the more conservative treatment with smaller doses and the experience in Japan tends to support the recommendation of doses of 1-2 IU/kg/day and a total ACTH dose of approximately 50 IU/kg.

METABOLIC DISORDERS

PEROXISOMAL DISORDERS

The total fatty acid and aldehyde composition in the brain, liver, and kidneys of two infants with Zellweger's syndrome and one with pseudo-Zellweger's syndrome and the fatty acid patterns expressed as percent values are reported from the Autonomous University of Barcelona, Hospital Infantil Vall d'Hebron, Barcelona, Spain. In confirmation of previous findings, patients with Zellweger's syndrome had extremely low levels of docosahexaenoic acid in the brain, liver, and kidneys. In both Zellweger's and pseudo-Zellweger's syndrome the ratio of the polyunsaturated fatty acids 22:6w3/22:4w6 was markedly decreased in all tissues. The findings reinforced the hypothesis of an enzymatic defect in peroxisomal disorders involving the desaturation of long polyunsaturated fatty acids. (Martinez M. Severe deficiency of docosahexaenoic acid in peroxisomal disorders: A defect of delta 4 desaturation? Neurology August 1990; 40:1292-1298).

COMMENT. In an excellent review of peroxisomal disorders (Naidu S, Moser HW. Neurologic Clinics August 1990; 8:507. W. B. Saunders Company, Philadelphia) the clinical signs of Zellweger's syndrome and other group I peroxisomal disorders are listed as follows: dysmorphism, hypotonia and retardation, early onset seizures, sensorineural hearing loss, retinal pigmentary degeneration, cataract, hepatomegaly. The biochemical and morphologic abnormalities include plasma increased very long chain fatty acids, phytanic acid, pipecolic acid; RBCs reduced plasmalogens, x-ray bony stippling, MRI central demyelination, liver absent peroxisomes, fibrosis and cirrhosis; kidney renal cortical cysts. Dietary treatment which effectively reduces plasma VLCFA levels is now available and bone marrow transplant has been partially effective in two patients.

HEADACHE

MIGRAINE AND CEREBELLAR ATAXIA

A four year old boy with migraine associated with focal cerebral edema, CSF pleocytosis, and progressive cerebellar ataxia is reported

from the Departments of Neurology and Radiology, Yale University School of Medicine, New Haven, CT. The onset was at one year of age with an episode of unresponsiveness which resolved within one day. At three years of age he suffered acute cortical blindness for 36 hours. At four years of age he presented with a pulsating left-sided headache and an acute right hemiparesis. An MRI showed increased signal in the left parietooccipital region, enhanced with Gadolinium. The next day he had a right focal seizure lasting five minutes and a seizure occurrence with persistent nonresponsiveness and right hemiparesis. On recovery over several days, the hemiparesis fluctuated, a left-sided headache resolved, but cerebellar ataxia developed and persisted. The CSF was clear with 60 WBC, protein 12 mg/dl, and glucose 75 mg/dl. A course of Propranolol has prevented recurrence of hemiparesis and headache but lower extremity spasticity and gait ataxia have persisted. (Goldstein JM, Shaywitz BA et al. Migraine associated with focal cerebral edema, cerebrospinal fluid pleocytosis, and progressive cerebellar ataxia: MRI documentation. Neurology August 1990; 40:1284-1287).

COMMENT. A so-called meningitic migraine with cerebral edema and autosomal dominant cerebellar ataxia has been reported previously in an adult (Fitzsimons RB, Wolfenden WH. Brain 1985; 108:555) but the present case is the first report in a child and with MRI documentation of a transient lesion. Severe prolonged migrainous symptoms and prolonged partial status epilepticus are characteristic features of the MELAS syndrome or mitochondrial encephalomyopathy which should be considered in the differential diagnosis of patients with headache and seizures. (Montagna P et al. Neurology 1988; 38:751).

Dr. John Wilson and his colleagues at the Hospital for Sick Children, Great Ormond Street, London, have emphasized the importance of food intolerance and an allergic mechanism for migraine in children. As a visitor at Great Ormond Street for one year I was impressed with the results of hypoallergenic dietary treatment for migraine in children, and the benefits of this approach to treatment as a substitute for drug therapy have been confirmed in my pediatric neurology practice. The short term elimination of cow's milk, egg, wheat cereals, chocolate, orange and cheese in the diet of the above child might be considered as a complement or substitute for the propranolol therapy.

Hockaday JM is the editory of a new book on "Migraine In Childhood" (Butterworth, Guildford, England 1989) which has received an excellent review by Dr. Richard O. Robinson (Dev Med Child Neurol 1990; 32:748).