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