
COMMENT. The authors felt that this clinical picture may represent the most severe end of the spectrum of fetal abnormalities attributable to phenytoin and/or sodium valproate. The anticonvulsant drug dosages taken by the mother were phenytoin 175 mg and sodium valproate 1 gram daily but the serum levels were not reported.

ACITH IN INFANTILE SPASMS

The relationship between dose of ACITH and the initial effect and long-term prognosis was investigated in 41 children with infantile spasms at the Department of Pediatrics, Kyoto University, Kyoto, Japan. ACITH therapy began at 2-13 months of age (mean: 11.8 months) and the average treatment lag was 4.5 months. All patients were treated with Vitamin B6, valproate, and other anticonvulsants without benefit before ACITH therapy was begun. All patients had hypsarrhythmia on the EEG. The cause was acquired in 30 patients, unknown in 8 and associated with developmental delay, and idiopathic in 3 patients whose development was normal. The doses in 16 patients was 0.5 mg (20 IU) for those older than one year of age and 0.25 mg (10 IU) for those less than one year; daily injections were given for two weeks, every other day for two weeks, and twice weekly for a total of 30 injections. One-half of these doses were used in 14 patients and doses were calculated on the basis of body weight (0.01-0.015 mg) (0.4-0.8 IU)/kg in 11 patients. More than 0.015 mg (0.6 IU)/kg/day of ACITH was needed for a good initial response of seizures and EEG abnormalities. Doses lower than 0.015 mg/kg/day provided less seizure control at the end of treatment and less EEG improvement. ACITH in a dose of 1.6-2.4 IU/kg/day and a total dose of 44-60 IU/kg resulted in better mental development than smaller doses, but side effects increased with larger daily doses or larger total doses. Seizures were controlled in 71-89% with doses above 0.6 IU/kg/day. ACITH-induced cerebral atrophy increased with the total dose of ACITH but was not seen when daily doses were 2.4 IU/kg/day or less and total dose was 60 IU/kg. (Ito M et al. ACTH therapy in infantile spasms: Relationship between dose of ACTH and initial effect or long-term prognosis. Pediatr Neurol July/Aug 1990; 6:240-244).

COMMENT. The authors recommend that ACTH doses based on body weight or body surface area should be used in future studies to compare the results obtained at different institutions. The doses used in this study from Japan were relatively small and ranged from 7.3-47.6 IU/m²/day whereas those recommended in some recent publications have been much higher and usually 150 U/m²/day. Snead OC (Pediatr Neurol May/June 1990; 6:147) advocated 150 U/m²/day for one week, 75 U/m²/day in the second week, and 75 U/m² on alternate days for the third week. Bobele GB, Bodensteiner JB from the University of Oklahoma and Morgantown,
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 22:6n3/22:4n6 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, phytic acid, pipolic 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