deficiency of holocarboxylase synthetase and 2. late infantile disease due to biotinidase deficiency. Neurological manifestations are prominent in the late-onset group and seizures may precede the cutaneous eruption and loss of hair.

Having encountered one such patient who presented at the age of 5 years with ataxia and seizures refractory to medications (In Nutrition, Diet, and your Child's Behavior. Charles C Thomas, Springfield, 1986.), I now make it a practice to prescribe biotin 10 mg daily as a therapeutic test when this diagnosis is suspected. Treatment reverses the organic aciduria so that a urine collection for analysis should precede administration of the vitamin. A dramatic response to a vitamin in a single daily dose is certainly an improvement over long-term anticonvulsant drug therapy with its attendant potential side-effects. Supplies of biotin from Roche Labs are available only for research at present (See Biotin. Ed. by Dakshinamurti K, Bhagavan HN. Ann. N.Y. Acad Science. New York, 1985; 447, 222-224, 297-313)

TUBEROUS SCLEROSIS AND INFANTILE SPASMS

Forehead plaques, smooth patches of slightly raised skin with a reddish or yellowish discoloration, can be the earliest skin manifestation of tuberous sclerosis (TS) according to the authors who describe 2 patients seen at Bath and Bristol, UK., presenting with infantile spasms at 3 and 5 months of age. (Fryer AE et al. Arch Dis Child 1987; 62:292-293)

COMMENT: Early diagnosis of tuberous sclerosis (TS) is important for genetic counselling and prognostic predictions. The prevalence of TS in patients with infantile spasms has been estimated at 25% or higher in some series. A Wood's light examination of the skin for hypopigmented maculae, a more frequent characteristic dermatologic manifestation of TS, is important in all infants with myoclonic spasms and hypsarrhythmia.

THYROTROPIN-RELEASING HORMONE (TRH): AN ALTERNATIVE THERAPY FOR INFANTILE SPASMS

Pediatric neurologists at the Central Hospital, Aichi Prefectural Colony, Kasugai, Aichi 480-03, Japan, compared the effects of TRH in 31 children and ACTH in 33 with severe epilepsy. Approximately half the cases had infantile spasms and the remainder had Lennox-Gastaut syndrome. In the TRH group, complete control of infantile spasms occurred in 7 of 13 (53.7%) and marked improvement of the EEG's was observed in 8 (61.5%). In the ACTH group, infantile spasms were controlled in 75%. TRH treated patients had no serious side-effects whereas 66.7% of the ACTH group had complications, including pneumonia, hypokalemia, cataracts, and brain shrinkage.

TRH-tartrate (TRH-t), 0.5 - 1.0 mg, was administered intravenously to determine immediate effects on seizures and EEG and then intra-muscularly once daily for 1 - 4 weeks. TRH was effective in controlling infantile spasms within 4 - 16 days of its initiation. Three of the 7 responders remained seizure-free for > 6 months. (Matsumoto A, Kumagai T, Takenchi T, Miyazaki S, Watanabe K. Epilepsia 1987;28:49-55)