

## Clinical Variation of Autoimmune Polyendocrinopathy-Candidiasis-Ectodermal Dystrophy in 68 Patients

This relatively rare entity, also called polyglandular endocrinopathy type I (PGE-I), is characterized by at least 2 of the following: hypoparathyroidism, hypoadrenalism, and mucocutaneous candidiasis. Other autoimmune diseases are often associated with this basic triad, including alopecia, pernicious anemia, gonadal failure, vitiligo, hypothyroidism or hyperthyroidism or chronic lymphocytic thyroiditis, and hepatitis. Ahonen et al have analyzed the interrelationships of these entities, as well as unguis dystrophy (pitted nails), steatorrhea, and keratopathy (Table).

Periodic malabsorption was observed in 12 patients, which was intensified with hypocalcemia, but some patients had steatorrhea when they were not hypocalcemic, and malabsorption preceded hypoparathyroidism in some. Neither keratopathy, pitted nails, nor enamel hypoplasia correlated with hypoparathyroidism or hypocalcemia, suggesting strongly that these were independent entities.

With the exception of candidiasis, none of these entities was manifest before the age of 12 months. Although most of the organ diseases occurred in childhood, some patients developed autoimmunity of some organs, including hypoparathyroidism, as adults. Interestingly, patients who developed Addison's disease as the first disease other than candidiasis tended to develop far fewer associated diseases.

Ahonen C, Myllarniessi S, Sipila I, et al. *N Engl J Med* 1990; 332:1829.

**Editor's Comment**—*This report greatly augments previous data on patients with PGE-I. The association of diseases and their time of appearance is surprising to me in that keratopathy and hypoplastic enamel are apparently not related to hypocalcemia or hypoparathyroidism. The relatively good prognosis of patients with only candidiasis and Addison's disease is worth noting. Unfortunately, the presumed autoimmune process in the gut that causes steatorrhea remains an enigma. Hypocalcemia, when present, needs to be controlled to minimize the*

*steatorrhea. Interestingly, in the patients with Addison's disease, the aldosterone deficiency was evident before the cortisol deficiency in approximately 50% of the cases.*

*The authors have published several previous reports of this syndrome, concerning mode of inheritance, oral findings, diagnosis and staging of hypocortisolism in progressive autoimmune adrenalitis, effective use of ketoconazole against candidiasis, the presence of adrenal and steroidal cell antibodies in evaluating risk of adrenocortical and ovarian failure, and the expression of PGE-I in association with human leukocyte antigen (HLA)-A, but not HLA-DR. The interested reader will find these and other pertinent references in the extensive bibliography of this article.*

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