Pachyonychia Congenita Project

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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.

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Failure of etretinate therapy in pachyonychia congenita

MADAM, We describe here the failure of etretinate treatment in a child with pachyonychia congenita. Lesions of pachyonychia usually persist for life and treatment is directed towards relief of the hyperkeratosis by topical keratolytic agents or oral vitamin A or both. Oral retinoids have been reported to be effective not only in the treatment of severe keratinization disorders but also in pachyonychia congenita.[1]

A 10-year-old girl, daughter of unrelated parents, had hyperkeratotic patches with well defined borders on the palms and soles with hyperhydrosis. Her fingernails and toenails were thickened and the undersurface was filled with a yellowish hyperkaratotic material (Fig. 1). Examination was otherwise normal. Laboratory investigations showed normal values for: CBC, ESR, SGOT, SGPT, and serum vitamin A level. Mycological examination of nails and skin scrapings was negative. Skin biopsy showed no abnormality except hyperkeratosis.

The patient was treated orally with etretinate (Tigason) 1 mg/kg/day. After 10 days of therapy excessive skin dryness, erythema, scaling and fissuring of the lips and perioral area, and glossitis with a burning sensation occurred and the dose was decreased to 0·5 mg/kg/day. Since there was no improvement in 5 weeks, the therapy stopped. In contrast to the report by Schönfeld[1] we failed to demonstrate any beneficial effect from 5-week course of etretinate therapy.

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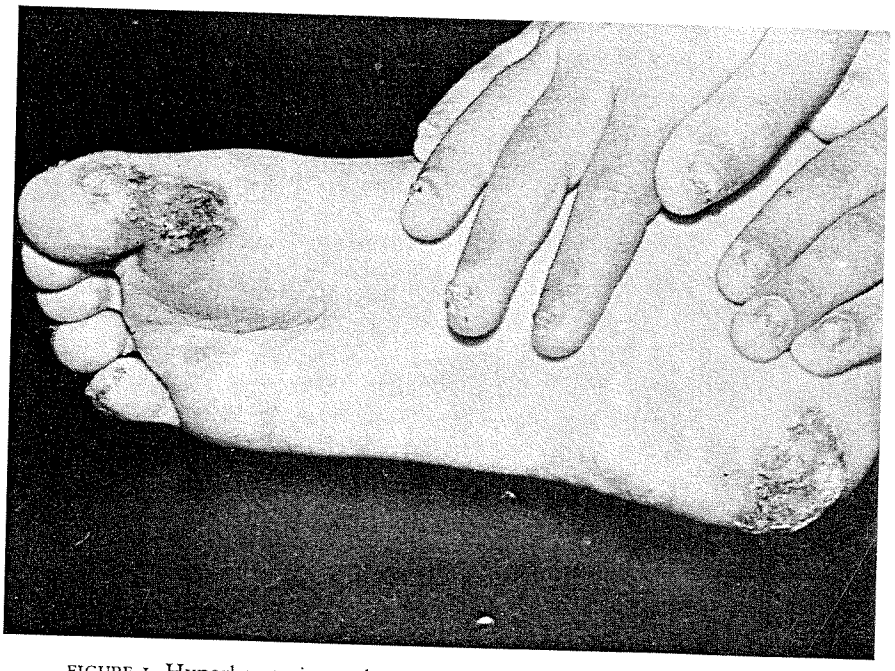

FIGURE 1. Hyperkeratotic patches on the sole and thickening of fingernails.

REFERENCE

1 Schönfeld P.H. The pachyonychia syndrome. *Acta Dermatovenereol* 1980; **60:** 45.