DISCUSSION

This study demonstrates a promising effect of short-wave ultraviolet light (UVB) on allergic contact dermatitis of the hands. Two out of the 3 patients where the eczema did not clear completely were treated for a relatively short period of time. They stopped treatment for personal reasons.

Two of the patients had a dyshidrotic-type eczema and one of these healed, while the other showed improvement in the contact dermatitis. Christensen (4) recently drew attention to the poor prognosis of dyshidrotic-type eczema of the hands in relation to nickel and the therapeutic difficulties.

In our study of the effect of UVB on delayed hypersensitivity in the guinea pig we showed that the suppression of cell-mediated reactivity was confined to the UV-exposed skin (2). Thus the effect of UVB is mainly local, and we have therefore not repeated the patch testing after the UVB treatment.

The mechanisms of action of UVB on the allergic contact dermatitis are not completely known. UV light has been shown to affect immunocompetent cells in different ways. Langerhans cells are important in the afferent phase of the immune response by presenting the antigen to immunocompetent lymphocytes. Small doses of UVB damage the surface markers of these cells (1). In addition, epidermal cells have an impaired antigen presenting function in UVB-irradiated skin (9). The inflammatory cells in tissue sections of allergic contact dermatitis consist of about 75% T-lymphocytes (8). The number of effector cells is regulated by suppressor T-lymphocytes which play a central role in controlling the immune response. T-lymphocytes are sensitive to UV-light (6) which affects both their functional capacity and their viability. All this indicates that UVB radiation reduces the number of effector cells both directly and indirectly through suppressor T-lymphocytes and thereby alleviates the allergic reaction.

Our conclusion is that UVB may be tried as a supplementary treatment for patients with long-standing allergic contact dermatitis where topical therapy has proved unsuccessful.

ADDENDUM

Three months after these results were recorded and summarized (Table I), 5 of the patients were treated once every second week with a UVB dose of 1.2 J/cm². There have been no relapses during this period of time.

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Treatment of the Ichthyosis of the Sjögren-Larsson Syndrome with Etretinate (Tigason®)

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Received April 7, 1982

Abstract. The ichthyosis of seven patients with the Sjögren-Larsson syndrome was treated with an aromatic retinoid, etretinate, during six months. Very good results were registered in six of the patients measured both as clinical improvement and as reduction in quantity of emollients needed. No unexpected side effects were noted.

Key words: Etretinate; Congenital ichthyosis; Sjögren-Larsson syndrome

Table I. *Etretinate treatment of SLS patients for 6 months*

The results of the therapy are measured as the decrease in the need for topical treatment from the prestudy period to the end of the test period, as well as a clinical assessment

Pat.	Age (yrs)	Weight (kg)	Etretinate, mg/day: Weeks 1–2	3–6	7–	Maintenance	Carbamide cream, g/wk: Prestudy	End	No. of baths/week: Prestudy	End	Clinical assessment
I.A.	74	56	50	25	10	10	150	15	7	7	Very good
R.S.	45	57	50	25	10	25	20	0	2	1	Very good
N.H.	42	45	50	25	10	25	50	25	7	7	Very good
E.L.	25	39	50	25	10	10	100	40	7	7	Good
T.J.	24	30	25	17.5	10	20	30	10	3	1	Very good
A.Å.	21	42	50	25	10	25	50	10	3.5	0	Very good
I.G.	18	39	50	25	10	25	35	15	14	7	Very good
							62	16	6.2	4.8	

Etretinate (Tigason®) has been reported to be of value in the treatment of various disorders of keratinization. Good results in the treatment of certain ichthyoses (lamellar, x-linked, i. vulgaris) have been reported, but the response of epidermolytic hyperkeratosis was considered less satisfactory (for ref., see 6).

The Sjögren-Larsson syndrome (SLS) in its fully developed form consists of mental retardation, spastic di- or quadriplegia and ichthyosis. The

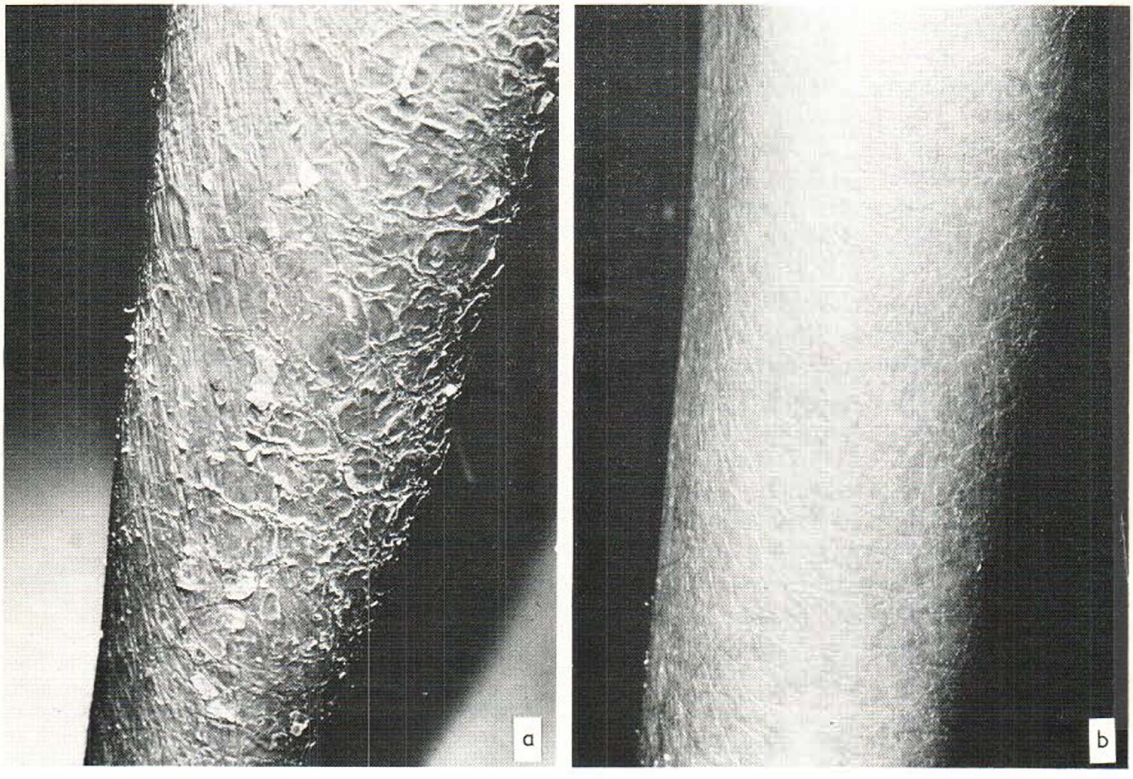

Fig. 1. Lamellar desquamation in a patient with the Sjögren-Larsson syndrome (*a*) before treatment with etretinate, (*b*) after treatment for one month.

ichthyosis is considered to be a variant of lamellar ichthyosis (2). In its adult form it has certain features distinguishing it from other ichthyoses (4) and ultrastructural differences in the stratum corneum have also been suggested (1). The SLS is a rare syndrome, with autosomal recessive inheritance. It has been reported sporadically from various parts of the world (7, 9). A certain clustering of cases is found in the northern parts of Sweden (3). This fact made it possible for us to study the effect of etretinate on the ichthyosis of the SLS in a series of patients.

PATIENTS AND METHODS

Seven patients with SLS were included in the study. The diagnosis of SLS is defined in the original work of Sjögren & Larsson (8). Etretinate was administered for 6 months, as shown in Table I. The initial daily dosage was about 1 mg per kg body weight during the first 2 weeks. During the following 4 weeks the dosage was reduced to about 0.5 mg/kg/day. From the seventh week a further reduction was attempted. When the state of the skin worsened, the dosage of etretinate was adjusted, eventually reaching the maintenance level shown in Table I.

A global assessment of the result of the treatment was made using the arbitrary categories very good, good, unchanged, and worse. In addition, the intensity of treatment was assessed for 2 weeks before the start of the study and then monthly. This was done by registering the number of baths and the amount of emollients used each week. All the patients had used the same emollient as the topical mainstay for a long time: carbamide 10%, water 20% in ung. Merck® (a vehicle with properties intermediate between ointment and cream).

The following laboratory tests were performed at the start of the treatment and then once a month: hemoglobin, red blood cells, hematocrit, MCV, MCH, MCHC; white blood cells and differential counts; thrombocytes; urinalysis; bilirubin, alkaline phosphatases, serum glutamyl transferase, aspartate-amino- and alanine-aminotransferase; serum electrolytes including calcium and phosphorous; serum cholesterol, and triglycerides.

RESULTS AND DISCUSSION

An increased desquamation of the ichthyotic skin was noted during the second week, reaching a maximum during the third week of treatment with etretinate. During this time there was an increase in pruritus in all patients, often leading to excoriations. The pruritus disappeared when the dosage was reduced. Already after 4 weeks all patients had a treatment result considered to be very good (Fig. 1). On reduction of the dosage the ichthyosis gradually increased. This made a successive increase to the maintenance level necessary. This level was chosen so as to produce a substantial improvement in the ichthyosis.

The influence of etretinate on the treatment intensity is shown in Table I. The amounts of carbamide cream necessary to relieve dryness were reduced in all cases. The decrease in the number of baths is less pronounced. This measure is less valid than the amount of topical treatment used, as most of the patients take daily baths as part of their daily hygienic routine and not only as a way of reducing the ichthyosis. On the whole the care of the skin of the patients was facilitated, i.e. became less time-consuming. This was highly appreciated, as the heavy daily skin care routine is very demanding. It was surprising that many of the parents of the patients considered the ichthyosis to be the most serious part of the syndrome because it imposed such a continuous work load.

No abnormalities were registered in the laboratory data. Apart from pruritus in all and cheilitis in one patient during the initial phase, no other side effects of importance were noted. The usefulness of etretinate for chronic medication in this syndrome will thus depend on its toxicity. The possible adverse effect on the blood lipids seems to be the most serious risk known at present (5).

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