known that B27 frequency is lower in idiopathic ankylosing spondylitis patients from the Mediterranean area in comparison to patients from the North-European regions (10).

Therefore, the ethnic background seems to influence the linkage between psoriatic arthritis and distinct alleles of the major histocompatibility system.

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HLA-B 16 in Hailey-Hailey's Disease

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Malchus R, Marsch W C, Ehlers G. HLA-B 16 in Hailey-Hailey's disease. Acta Derm Venereol (Stockh) 1986; 66: 264–266.

Hailey-Heiley's disease is an autosomal hereditary disease of the skin for which only few data exist in regard to genetic markers concerning the HLA system. We report on HLA-A, B and C typing results finding an increased frequency of 55.5% HLA-B 16 positive patients compared to 8.2% in healthy controls. *Key words: MHC; HLA-B locus.* (Received December 23, 1985.)

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Hailey-Hailey's disease (chronic benign familial pemphigus) is an autosomal dominant hereditary cutaneous disease with irregular penetrance (1, 3). The disorder is based on a defect of intercellular coherence of the epidermal keratinocytes. The histological hallmark is a suprabasal blister formation with acantholytic keratinocytes. Clinically the disease is manifested by physical and chemical insults and preferentially affects the intertriginous areas. However, the disease can be expressed in a generalized fashion (5).

It required six years to gather ten unrelated patients. Since there seems to be a predisposition in sibs of the affected we looked for possible associated HLA antigens. Only one HLA study has been performed until now showing a normal distribution of the HLA antigens (4).

MATERIALS AND METHODS

10 patients and 1 267 unrelated healthy blood donors were HLA typed (microlymphocytotoxicity test, NIH standard technique (2)) using up to 180 specific HLA antisera allowing discrimination of HLA-A 1, 2, 3, 11, 23, 24, 25, 26, 28, 29, 30, 31, 32, 33, B 7, 8, 13, 14, 18, 27, 35, 37, 38, 39, 41, 44, 45, 47, 49, 50, 51, w52, w53, w54, 55, 56, 57, 58, 60, 61, 62, 63, Cw 1, 2, 3, 4, 5, 6, and 7. For statistical calculations we used the chi-square test and corrected the p value by multiplication with the number of comparisons, i.e. the number of antigens tested.

RESULTS

Eight male and two female patients suffering from Hailey-Hailey's disease were typed for HLA from 1978 to 1984. Patient 7 was excluded from statistical evaluation because of his Arab origin. Five out of nine European patients were typed HLA-B 16 positive versus 104 out of 1 267 healthy individuals (see Table I, chi-square=25.6 $p<0.0001$, $p_c<0.003$). No significant differences were seen for any of the other antigens tested.

The mean age of manifestation of the disease was 34 years in the HLA-B 16 positives vs. 41 years in the HLA-B 16 negatives. Also no relevant difference between the HLA-B 16 positives and negatives was seen in regard to affected relatives.

DISCUSSION

HLA-B 16 consists of two splits (HLA-B 38, HLA-B 39) and the group of HLA-B 16 positives is reconstituted from the HLA-B 38 and HLA-B 39 positives. Actually the increased frequence of HLA-B 16 in patients is due to the increased frequence of HLA-B

Table I. *Distribution of sex, age of manifestation, HLA antigens and affected relatives in ten cases of Hailey-Hailey's disease*

Case no.	Sex	Age of manifestation in years	HLA-A	HLA-B	Cw	HLA-B 16[a]	Affected relative
1	Male	23	2, 25	38, 62	3, –	+	Mother, son
2	Male	23	2, 31	7, 39	–, –	+	No
3	Female	39	2, –	14, 39	–, –	+	No
4	Male	42	1, 2	13, 38	–, –	+	No
5	Female	43	2, 24	44, 39	–, –	+	Unknown
6	Male	11	1, 28	8, w52	7, –	+	Father, brother
7[b]	Male	27	11, –	51, –	4, –	–	Mother
8	Male	35	1, 26	7, –	–, –	–	No
9	Male	39	2, 31	51, –	–, –	–	Brother
10	Male	78	3, 25	8, –	–, –	–	No

[a] HLA-B 16 positive individuals are either positive for HLA-B 38 or HLA-B 39. The frequency of HLA-B 16 positive patients (5 out of 9=55.5% is significantly different compared to healthy controls (104 out of 1.267=9.2%), chi-square=25.6, $p<0.0001$, $P_c<0.00.3$.

[b] Case 7 has been excluded from statistical calculations for its Arabic origin.

39, 44% vs. 3.3% in our controls. But since the frequence of HLA-B 38 was 11% vs. 3.5% in the controls (no significant difference), we thought it justified to refer to an increase in HLA-B 16.

It is always problematic to apply statistical methods to data of only few patients and even more dubious to draw conclusions on such evaluations. Hailey-Hailey's disease is a fairly rare disease, however, the pathomechanism of which could be easier assessed if all investigated cases were published.

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Circulating Lymphocyte Subsets in Patients with Alopecia areata

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Baadsgaard O, Lindskov R. Circulating lymphocyte subsets in patients with alopecia areata. Acta Derm Venereol (Stockh) 1986; 66: 266–268.

Lymphocyte subsets in peripheral blood of fourteen patients with patchy alopecia areata or alopecia universalis were estimated using monoclonal antibodies and immunofluorescence. The median percentage of circulating Leu 2a, 3a, 4 and 7 positive cells ("T-suppressor/cytotoxic", "T-helper/effector", total T-cells and killer and natural killer cells) were normal.
Key words: T-lymphocytes; Killer and natural killer cells. (Received March 14, 1985.)

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In spite of conflicting findings regarding cell-mediated immunity and auto-immune phenomena in alopecia areata patients, there is evidence indicating an immunologic abnormality as an etiological factor (1, 2, 3, 4, 5). On this background we have measured circulating lymphocyte subsets in patients with alopecia areata.

MATERIALS AND METHODS

Patients

Fourteen consecutive outpatients, 9 females and 5 males, between 16 and 71 years of age (median 32), participated in the investigation. Eleven patients suffered from patchy alopecia areata (AA) and 3