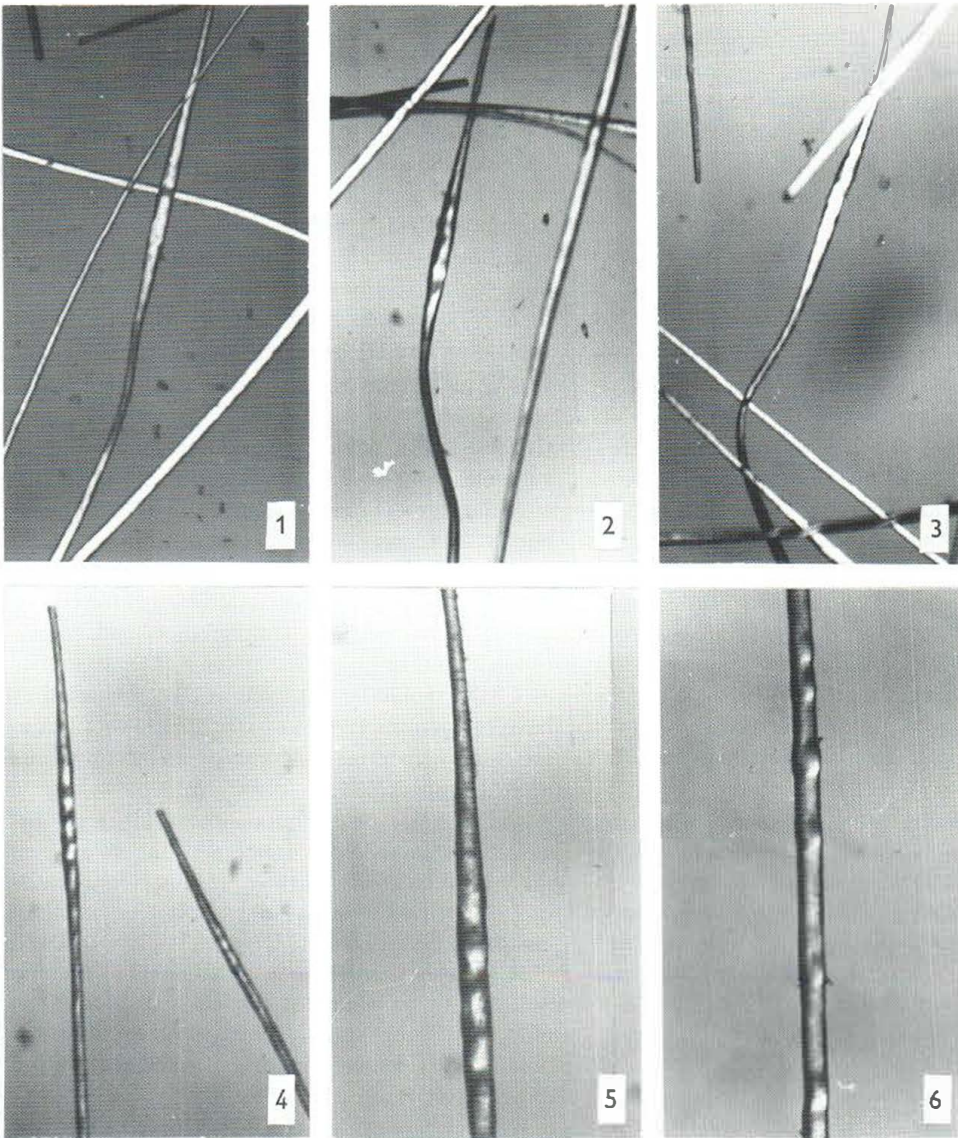




The Hair in Acrodermatitis Enteropathica—a Disease Indicator?

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Received September 13, 1978

We have had under observation an infant boy, aged 9 months, with a typical acrodermatitis enteropathica (A. E.): eczematiform and psoriasiform skin

Figs. 1, 2 and 3. The ends of the hair are "spearhead-like" with a general swan-neck appearance.

Figs. 4, 5 and 6. The hair displays numerous striae, with a slight trichonodosis.

lesions around the body orifices, mucosal lesions, paronychia, widespread and almost total alopecia, association with gastrointestinal malfunction and low zinc levels. Zinc sulphate therapy was instituted and within 10 days the whole situation had improved and the hair had grown.

The hair itself was carefully examined. It was white, sparse, short (less than 1 centimetre), very brittle, and looked like down. On optical light microscopic examination, the hair did not show any evident anomaly, but only slight, non-specific changes. On the other hand, we were surprised to find very characteristic anomalies with the polarizing microscope; indeed, examination with polarized light revealed a very specific defect, not reported previously, including the following numerous and simultaneous aberrations: (1) the ends of the hair are spindly, moniliformis, sharp at the distal end, "spearhead-like"; (2) at the proximal end of the spindle, there is a narrow and deficient area which gives rise to an elegant wave; (3) sometimes there is another elegant wave, lying in the opposite direction, thus giving a general swan-necked appearance; (4) furthermore, the whole shaft displays numerous striae, disposed in little groups of three or four, which correspond to areas of brittleness—a kind of mild trichonodosis, or slight trichoschisis.

The juxtaposition of these very strange aberrations probably renders the patient's hair a specific hair defect, an indicator of the disease, directly related to zinc deficiency. Such findings have not been reported previously.

Successful results were obtained with zinc therapy. Within a month the eruption resolved, the diarrhea stopped and the growth of hair was gradually restored. Three months later a new examination with polarized light showed a quite normal structure of the hair shafts: all the previously observed anomalies had disappeared.

Chronic Bullous Dermatitis of Childhood

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Received August 22, 1978

Abstract. A case of chronic bullous dermatitis of childhood in a 3-year-old boy is described. Immunofluorescence tests were negative and biopsy of the jejunal mucosa showed marked villous atrophy. The dermatosis

was brought under control by a combination of diamino-diphenylsulphone and systemic steroids. The relationship with other bullous eruptions of childhood such as dermatitis herpetiformis and bullous pemphigoid is discussed.

The chronic acquired bullous dermatoses of childhood include: pemphigus vulgaris, bullous pemphigoid (BP) and dermatitis herpetiformis (DH), with their typical clinical, histological and immunological features. Moreover, various cases of bullous dermatosis have been described, having a clinical picture almost exclusively of large blisters, with a histological picture of subepidermal bullae and generally with negative immunofluorescence (IF) tests, responding variably to diaminodiphenylsulphone (DDS) or sulphapyridine. For this disease the name chronic bullous dermatosis of childhood (CBDC) was proposed (7).

However, some authors prefer to use the term juvenile DH (5, 2) even in the absence of the typical immunological findings. On the other hand, review of the literature suggests that some cases described as CBDC should be classified as DH, because they have IF features typical of DH: deposits of IgA at the dermo-epidermal junction or at the tip of the dermal papillae (1, 8).

As far as intestinal abnormalities in blistering diseases of childhood are concerned, the atrophy of the jejunal mucosa, which is present in most cases of DH (3), was found in cases described as BP of childhood (9), or juvenile DH (10); in these cases IF tests were not carried out. Other authors found a non-specific inflammatory reaction in the jejunal mucosa (5), or abnormalities of gastrointestinal functions (2) in some patients with a clinical picture of juvenile DH.

We now report a case of CBDC in which it was possible to perform the biopsy of the jejunal mucosa and the IF tests.

CASE REPORT

A 3-year-old boy was admitted to our Department in July 1977 with an extremely pruritic widespread bullous dermatosis, which started in January 1977. The initial lesions—vesicles and blisters with secondary erosions and crusts—appeared on the pinna. Subsequently lesions appeared on the lower extremities, buttocks, penis and abdomen. The dermatosis ran a chronic course. In May 1977, after admission to a local hospital, a diagnosis of juvenile pemphigoid was made on the basis of clinical data and a Tzanck smear test. He was treated with sulpha-