

Incidence and Prevalence of Dermatitis herpetiformis in Western Sweden

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The annual incidence of IgA-positive dermatitis herpetiformis (DH) during the years 1976-1981 in Gothenburg, Sweden, a city with approximately 430 000 inhabitants, was estimated prospectively. The mean annual incidence was 1.1 per 10⁵ inhabitants. The prevalence of DH as at 31 December, 1981 was 22.9 per 10⁵ inhabitants or 19.6 if patients in long-standing spontaneous remission were excluded. These figures probably represent the minimum overall prevalence rate. *Key words: Dermatitis herpetiformis; Epidemiology; Incidence; Prevalence.* (Received November 19, 1983.)

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Dermatitis herpetiformis (DH) is a chronic pruritic and blistering disease associated with a gluten-sensitive enteropathy. This disease seems to be present in all parts of the world, but epidemiological data are scarce. A study of the incidence of DH in Finland showed that 1.3 persons per 100 000 inhabitants are estimated to contract DH annually (1). Only a few reports on the prevalence of DH have been published, with divergent results. Thus, in England a minimum of 1.2-1.4, in Northern Ireland 2.5 and in Finland 10.4 persons per 100 000 inhabitants have been estimated to suffer from the disease (1, 2, 3). DH is rare in Japan (4).

It is not known to what extent the geographical variation in the occurrence of DH is explained by differences in epidemiological methods, diagnostic criteria or genetic and environmental conditions for the development of DH. Previous reports have been based on records for patients attending dermatological clinics. All patients are, however, not cared for by dermatologists. Since the clinical manifestations of DH vary inter- and intraindividually, patients with discrete symptoms may consult non-dermatologists and be unrecognised, those in remission may not consult at all and a few patients with severe malabsorption as the major symptom may be treated by gastroenterologists. It is therefore possible that a considerable number of patients with DH may remain hidden in the population not receiving dermatological care. In addition, the inadequacy of classical diagnostic criteria—clinical, histological and pharmacological—has been stressed (5). The occurrence of granular deposits of IgA in the papillary tips of perilesional as well as apparently normal skin is now generally accepted to be the most reliable diagnostic criterion of DH (6). The immunofluorescence method was not available or consistently used in the studies previously referred to. Furthermore, it has only recently been recognised that patients having a homogeneous linear deposition of IgA along the basement membrane zone represent a separate clinical entity—linear IgA dermatosis—and should not be included among the patients with granular deposition of IgA (7).

The aim of this study was to examine prospectively the occurrence of DH in a defined population in Sweden.

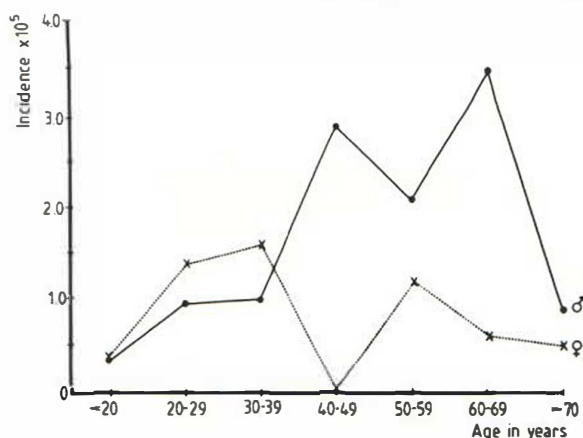


Fig. 1. Annual incidence (1976-1981) of dermatitis herpetiformis in males and females in different age-groups in Gothenburg, Sweden, calculated as a ratio of all residents of the same sex and age (mean values).

PATIENTS AND METHODS

Study area

Gothenburg is the second largest city in Sweden and is situated on the west coast. The population in 1976 and 1981 totalled 442410 and 428 171 inhabitants respectively, corresponding to about 5% of the Swedish population (8). The sex distribution shows a slight surplus of females. Foreign citizens make up 9.1% of the population of Gothenburg. The main occupations in the study area are manufacture, trade, communications and public services.

There is one university department of dermatology with 17 dermatologists, 4 smaller public outpatient clinics staffed by 1-2 dermatologists and 3 full-time private dermatologists. Most dermatologists meet twice a week for clinical rounds and postgraduate training.

Collection of patients

During the last 15 years we have had a special interest in DH and actively recruited patients from Gothenburg and the surrounding region. These patients attend a special outpatient clinic staffed by 2 dermatologists and one assistant nurse. An exhaustive search for patients with DH started in 1976. All dermatologists in the area were contacted personally and requested to refer all patients with known or suspected DH. The diagnostic indices of the university hospital and the 4 smaller dermatological clinics in the city were inventoried back to 1967. In addition, the patient records at the gastroenterological section of the Department of Medicine of the University of Göteborg were checked for patients with a combination of DH and malabsorption. These procedures were repeated several times during the following years. Since patients with onset of DH in 1981 may be detected after the deadline for this study (31 December, 1981) the patients with DH diagnosed during 1982 were questioned concerning onset of symptoms. Finally, it was checked that all patients from the city in whom biopsies for histopathological or direct immunofluorescence examination had resulted in the diagnosis of DH had been considered for inclusion in the study.

Methods

Examination of punch biopsies by means of the direct immunofluorescence technique has been performed in Gothenburg since 1976 (L.-Å. N.). Two punch biopsies (3 mm) were taken under local anaesthesia from normal skin of the lumbosacral region in all patients referred to the DH clinic, irrespective of previous examinations. The specimens were immediately frozen in liquid nitrogen. The direct immunofluorescence technique has been described elsewhere (9).

Table I. The year of onset of dermatitis herpetiformis in patients living in Gothenburg between 1976 and 1981

	1976	1977	1978	1979	1980	1981
Patients	5	6	4	5	5	4

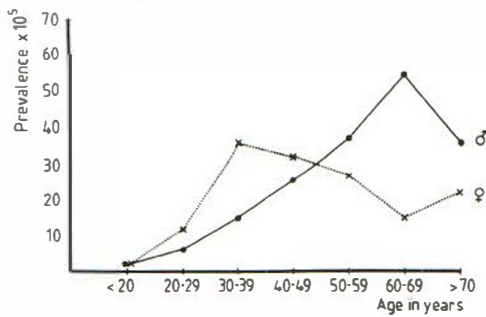


Fig. 2. Prevalence of dermatitis herpetiformis in males and females of different age-groups in Gothenburg, Sweden, as at 31 December 1981 as a ratio of all residents of the same sex and age.

In order for a patient to be included in this study, the clinical and histopathological suspicion of DH had to be supported by the presence of granular IgA in the papillary tips or along the basal membrane of uninvolved skin. The IF examination was repeated in all patients who had become free from symptoms without treatment. Four patients with linear homogeneous IgA along the basal membrane zone have been reported elsewhere and are not included in this study (9).

The patients were interviewed, examined and followed up at 3–6-month intervals. Most of them underwent gastroenterological and haematological investigations (10, 11).

RESULTS

Eighteen men and 11 women contracted DH during the six-year period 1976–1981 (Table I). The mean interval between the onset of skin symptoms and diagnosis as confirmed by IF was 8 months (range 1–24, median 6). The mean annual incidence, i.e. the annual number of patients from Gothenburg with onset of DH, was 1.1 per 100 000 inhabitants. The mean annual incidence in different age-groups is shown in Fig. 1. It is possible to contract DH at any age, but DH is rare in children. The peak rate for males occurred between 40 and 70 years, whereas the onset of the disease in females was evenly distributed throughout adult life.

There were 84 patients (43 males) with signs and symptoms of DH including those controlled by gluten-free diet, living in the City of Gothenburg on 31 December 1981. The overall prevalence rate of DH was 19.6 per 100 000 inhabitants. The mean age of onset was 37 years (range 5–79). On 31 December 1981 the mean duration of DH was 15 years (range 3/12–57, median 12). The age distribution of this population of DH cases is shown in Fig. 2. The number of patients with DH increases with age, as may be expected with a chronic disease.

Table II. A comparison between epidemiological data and presumed pathogenetic factors for dermatitis herpetiformis in two Scandinavian countries

Geographical area	Incidence per 10 ⁵ inhabitants	HLA-B8 (%)		Dietary gluten (g/day)	Dietary iodine (µg/day)	References
		Controls	DH			
Finland	1.3	17	87 ^a	20	220	Reunala et al. (17) Reunala & Lokki (1)
Gothenburg, Sweden	1.1	25	81 ^a	15	200–250	Present study Becker (18) Anderson et al. (19)

^a DH diagnosis confirmed by direct immunofluorescence (10).

Patients in natural remission (normal diet and no skin lesions or treatment for at least the last 6 months) were not included in the prevalence calculations. We are aware of 14 such patients (4 males), all of whom still had granular deposits of IgA in the skin. If they are included the overall prevalence rate of DH was 22.9 per 100000 inhabitants.

DISCUSSION

The annual incidence of DH in residents of Gothenburg for the last 6 years was estimated to be 1.1 cases per 100000 inhabitants. Figures from earlier periods are not included since other diagnostic criteria for DH were then used. There is reason to believe that the number of newly diagnosed cases of DH not included in this intensive 6-year search for cases in Gothenburg was negligible. The organisation of medical care in Gothenburg—with an ample number of dermatologists—and the fact that DH is a pruritic disease with a poor response to topical corticosteroids make it highly probable that a patient will consult a dermatologist at an early stage and finally be referred to the specialised outpatient clinic.

Since the number of patients is small, it is difficult to draw valid conclusions concerning the incidence of DH in the general population of Sweden. However, extrapolation of the incidence figures from Gothenburg gives about one hundred new cases per year in Sweden (95% confidence interval, 60–136). The incidence rate is consistent with that found in the carefully performed study in Finland in 1978 (1). This fits well with the observation that the genetic and environmental factors most commonly associated with DH—HLA-B8, DR3 and dietary gluten and iodine—seem to be similar in the two countries (Table II). Exact incidence figures are not known for countries where the above predisposing factors markedly deviate. However, DH is very uncommon in Japan, where HLA-B8 is rare (4). DH seems to be somewhat less common in England than in Scandinavia and the gluten intake is only about half as high (2, 3, 12). Population studies from different countries may help us to discover and evaluate factors contributing to our understanding of the disease. When regularly performed, such studies will also permit the early discovery of changes in the incidence of the disease.

The prevalence rate of DH in Finland was only half of that in the present study (1). This discrepancy may be explained by our inclusion of patients with only minor symptoms and patients with malabsorption as a dominant symptom and treated by gastroenterologists. We are aware of two brief reports on the prevalence of DH in Sweden. DH occurred in 29.8 persons per 100000 inhabitants in central Sweden (13) and concomitantly with adult coeliac disease in 19.8 per 100000 inhabitants above the age of 15 years in a district of eastern Sweden (14). It is therefore possible that the present study underestimates the prevalence of DH. Patients who have been in long-standing remission may be unknown to us. In fact, Fry et al. (15) have reported that 14% of DH patients on normal diet did not require drugs. In addition, patients who have been maintained for many years successfully on dapsone may receive refills from general practitioners. Finally, it cannot be excluded that a few patients have left Gothenburg possibly due to increasing unemployment.

It is interesting to compare these prevalence rates with the result of a recent epidemiological study in eastern Sweden of childhood and adult coeliac disease (16). The overall prevalence rate (unaccompanied by DH) was calculated to be 56/100000, and considered to be a minimum rate.

Knowledge of the total number of cases of DH is useful for planning the resources necessary for the adequate care of these patients. However, conclusions drawn from the data in this study may only be valid for populations which are comparable with respect to ethnic origin and socioeconomic levels to the one we have studied.

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