Acta Derm Venereol (Stockh) 1992; 72: 25–27

Coeliac-type Dental Enamel Defects in Patients with Dermatitis Herpetiformis

LIISA AINE[1], MARKKU MÄKI[2] and TIMO REUNALA[3]

[1]*Department of Oral and Maxillofacial Surgery, University Hospital of Tampere,* [2]*Department of Clinical Sciences, University of Tampere, and* [3]*Department of Dermatology, University Hospital of Tampere, Tampere, Finland*

The teeth of 30 adult patients with dermatitis herpetiformis and 66 sex- and age-matched healthy controls were examined for dental enamel defects. Sixteen of the patients (53%) with dermatitis herpetiformis, opposed to only one (2%) of the healthy controls ($p < 0.001$), were found to have coeliactype permanent-tooth enamel defects. The grades of these defects were milder than those described for severe coeliac disease. There was no correlation between the degree of enamel defects and jejunal villous atrophy. The present finding of frequent coeliac-type dental enamel defects in adults with dermatitis herpetiformis suggests that these patients were already suffering from subclinical gluteninduced enteropathy in early childhood, at the time when the crowns of permanent teeth develop. ***Key words: Gluten enteropathy; Hypoplasia.***

(Accepted July 25, 1991)

Acta Derm Venereol (Stockh) 1992; 72: 25–27.

L. Aine, Department of Oral and Maxillofacial Surgery, University Hospital of Tampere, SF-33520 Tampere, Finland.

Dermatitis herpetiformis (DH) is a chronic skin disease which can appear at any age. Small pruritic blisters, macules and papules can be detected, especially on the knees, elbows and buttocks. Patients present pathognomonic IgA deposits in the uninvolved skin and also in the oral mucosa (1, 2). DH is strongly associated with coeliac disease. The HLA pattern is similar (3) and most patients exhibit glutensensitive small-bowel villous atrophy (4). Elimination of gluten from the diet usually heals the rash (4), and a lifelong gluten-free diet is the treatment of choice for these patients (3).

In our previous studies we have shown that a high percentage of children (6) and adults (7) with coeliac disease have symmetrically and chronologically distributed dental enamel defects in all four sections of the permanent dentition. These developmental enamel defects must have formed in early childhood, at the time when the crowns of permanent teeth mature, that is, before the age of 7 years. These coeliac-type enamel defects also frequently occur in children with DH (8).

The present study investigated whether adult patients with DH have enamel defects similar to those found in patients with coeliac disease. Because patients with DH have jejunal abnormalities ranging from severe villous atrophy down to minor inflammatory changes, we correlated the jejunal and dental findings in our patients.

PATIENTS AND METHODS

Patients and controls

Thirty patients, 19 men and 11 women, attending a special outpatient clinic for DH patients at the Department of Dermatology, University Hospital of Tampere, were examined. The inclusion criteria were age (between 20–50 years) and the presence of full dentition (fewer than five teeth extracted and/or only one crowned tooth). The patients were studied at the ages of 25–50 years, the mean age being 37 years. The mean age at diagnosis of DH was 28 (range 16–42) years. The diagnosis was based on the clinical picture and on the demonstration of granular IgA deposits in the uninvolved skin by direct immunofluorescence examination (5).

Jejunal biopsies were performed at the time of initial diagnosis in 28 patients, and the mucosal findings were graded as subtotal villous atrophy (SVA, villous height less than 150 μm), partial villous atrophy (PVA, villous height 150 to 250 μm) and normal mucosa or slight changes (villous height greater than 250 μm) (9). Twenty-three patients were treated with dapsone and a gluten-free diet (GFD), five with GFD and two with dapsone alone. At the time of the dental examination, 11 patients on GFD were still using dapsone, but 12 had been able to stop the drug treatment.

The control group consisted of 66 healthy subjects (42 men and 26 women), chosen consecutively from a dental practice. Thus, all the patients with DH had at least two age- and sex-matched control subjects.

Dental examination

The enamel defects were clinically examined as previously described (6, 7). The colour, type and site of the defects were recorded, as were the number of fillings. The defects were classified as symmetrically and chronologically distributed, coeliac-type enamel defects, and as noncoeliactype defects. In the latter group, the lesions were on only one side of the dentition, the other side was intact. The chronology of the lesions also varied in this group.

The grade of the coeliac-type enamel defects was determined as

Table I. *Dental enamel defects in patients with dermatitis herpetiformis (DH)*

Enamel defects	DH (N=30)		Healthy controls (N=66)	
	N		N	
Coeliac-type defects	16	(53%)	1	(2%)*
Grade I	6		1	
Grade II	10		0	
Grade III–IV	0		0	
Nonspecific defects	14	(47%)	60	(91%)
No defects	0		5	(8%)

* $p < 0.001$ (Fisher's exact test)

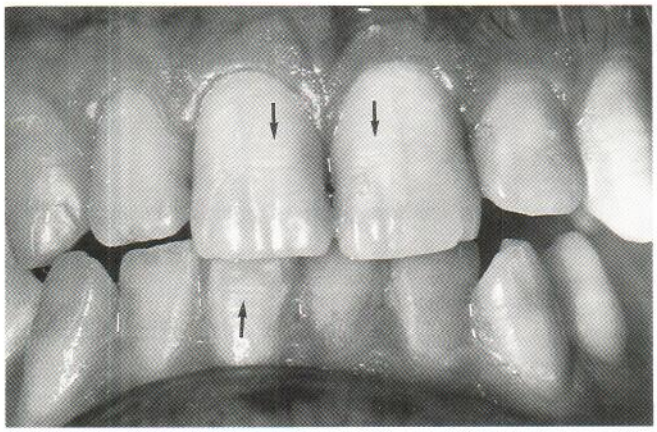

Fig. 1. Symmetrically and chronologically distributed coeliac-type permanent-tooth enamel defects of grade II in a 27 year-old male patient with dermatitis herpetiformis (DH). The typical horizontal grooves can be seen (arrows).

previously described (7). In brief, grade I enamel lesions include defects in the colour of the enamel; grade II slight structural defects with a rough enamel surface and horizontal grooves or shallow pits; grade III evident structural defects with part or all of the surface of the enamel rough and filled with deep horizontal grooves varying in width or with large vertical pits; and grade IV severe structural defects in which the shape of the tooth has also changed.

RESULTS

The prevalence and grade of the enamel defects found in the teeth of patients with DH are shown in Table I. Altogether 16 patients (53%) were found to have coeliac-type dental enamel defects, as opposed to only 2% of the healthy controls ($p < 0.001$). All the defects were mild, grade I or grade II lesions (Fig. 1). Unspecific noncoeliac-type defects were common in both DH patients (47%) and controls (91%). The difference between the study group and the controls was also clear when the total number of affected teeth were counted. Enamel defects were found in 407 (51%) of 793 teeth in DH patients, as opposed to 326 (18%) of 1,780 teeth in the control group.

Jejunal findings

Jejunal biopsy showed SVA in ten, PVA in fourteen and normal mucosa or slight changes in four patients. Three patients presented with minor gastrointestinal symptoms and one had a history of duodenal ulcer. None of the 30 patients had a history of symptoms suggestive of coeliac disease in childhood.

Coeliac-type enamel defects were found in 58% of the patients with SVA and PVA, and in one of the four patients with normal jejunal mucosa (Table II).

DISCUSSION

The present study clearly shows that adults with DH have symmetrically and chronologically distributed enamel defects similar to those described in patients with coeliac disease (6, 7, 10, 11). Coeliac-type dental enamel defects were found in 53% of our adult patients with DH. Previously, we demonstrated similar defects in 83% of 40 adults with coeliac disease (7). Adults with coeliac disease and the present DH patients did not exhibit the most severe, grade IV enamel defects which occurred in 11% of the children with coeliac disease (6).

The mechanism of coeliac-type systemic enamel defects remains obscure. The crowns of permanent teeth develop during the first seven years of life, and the disturbances which may cause developmental defects in enamel are thought to be nutritional or immunologic. Malabsorption and hypocalcaemia during enamel formation could be one reason for enamel hypoplasia (12). In support of this hypothesis, we have previously presented evidence that, in children with coeliac disease, a severe clinical presentation is correlated with more severe enamel defects (6). Another possibility could be that coeliac-type permanent-tooth enamel lesions are of immunologic origin and form an independent disease entity associated with gluten-sensitive diseases.

The coeliac-type enamel defects found in 53% of the present adult patients with DH suggest that these patients already had subclinical coeliac disease in childhood, i.e. a long time before they contracted their skin disease. However, none of the present adult patients or the DH children with enamel defects described previously (8) had any apparent clinical history suggestive of childhood coeliac disease. Children or adults with DH rarely present with short stature, which also argues against severe malabsorption and nutritional deficiences during their childhood (5, 13). However, subtle alterations leading to enamel hypoplasia may occur, and it is now becoming evident that the true prevalence of childhood coeliac disease seems to be much higher than previously thought (14). Many of these childhood cases present without any gastrointestinal complaints, although the small intestinal mucosa are thought to be affected early in life. These children may easily evade diagnosis and only develop clinical coeliac disease in adult life, or they may also contract DH with or without gastrointestinal symptoms.

Most of the present patients with DH had either subtotal or partial villous atrophy at the time of diagnosis. There seemed to be no correlation between the degree of mucosal damage and the presence of enamel defects. We also found one patient with grade II enamel defects who had only slight changes in the jejunal mucosa. However, we cannot exclude the possibility that this patient had had villous atrophy in childhood. It is well known that, after puberty, coeliac disease may remain

Table II. *Coeliac-type dental enamel defects and jejunal morphology in 28 patients with dermatitis herpetiformis (DH)*

Jejunal morphology	Number of patients	Enamel defect		
		Grade II N	Grade I N	Total N
Subtotal villous atrophy	10	4	2	6 (60%)
Partial villous atrophy	14	4	4	8 (57%)
Normal mucosa or slight changes	4	1	0	1 (25%)

latent and show normal villous architecture even though the patient continues on a normal diet (15). Increased amounts of dietary gluten or other triggering factors may then precipitate the disease, and this also seems relevant for some patients with DH who require increased amounts of dietary gluten to express jejunal villous atrophy (16).

The present finding of frequent but mild coeliac-type dental enamel defects in the permanent teeth of adult patients with DH suggests that several of the patients had been suffering from subclinical coeliac disease since early childhood. The same assumption seems valid for patients with adult coeliac disease, on the basis of both epidemiological (17, 18) and dental studies (7). At present the factors which trigger the gluten-sensitive skin disease, i.e. DH, in some but not all adult patients with long-lasting subclinical coeliac disease remain obscure. Further research is also needed to resolve the pathomechanism of coeliac-type dental enamel defects described in our present and previous studies (6–8).

ACKNOWLEDGEMENTS

This study was supported by the Emil Aaltonen Foundation and by the Tampere City Scientific Fund.

REFERENCES

1. Hietanen J, Reunala T. IgA deposits in the oral mucosa of patients with dermatitis herpetiformis and linear IgA disease. Scand J Dent Res 1984; 92: 230–234.
2. Nisengard RJ, Chorzelski T, Maciejowska E, Kytt L. Dermatitis herpetiformis: IgA deposits in gingiva, buccal mucosa and skin. Oral Surg 1982; 54: 22–25.
3. Hall RP. The pathogenesis of dermatitis herpetiformis. Recent advances. J Am Acad Dermatol 1987; 16: 1129–1144.
4. Kósnai I, Karpati B, Savilahti E, Verkasalo M, Bucsky P, Török E. Gluten challenge in children with dermatitis herpetiformis: a clinical, morphological and immunohistological study. Gut 1986; 27: 1464–1470.
5. Reunala T, Kósnai I, Karpati S, Kuitunen P, Török E, Savilahti E. Dermatitis herpetiformis: jejunal findings and skin response to gluten free diet. Arch Dis Child 1984; 59: 517–522.
6. Aine L. Dental enamel defects and dental maturity in children and adolescents with coeliac disease. Proceedings of the Finnish Dental Society 1986; 82(Suppl 3): 1– 71.
7. Aine L, Mäki M, Collin P, Keyriläinen O. Dental enamel defects in celiac disease. J Oral Pathol Med 1990; 19: 241–245.
8. Aine L, Reunala T, Mäki M. Dental enamel defects in children with dermatitis herpetiformis. J Pediatr 1991; 118: 572–574.
9. Reunala T, Blomqvist K, Tarpila S, Halme H, Kangas K. Gluten-free diet in dermatitis herpetiformis. I. Clinical response of skin lesions in 81 patients. Br J Dermatol 1977; 97: 473–480.
10. Smith DMH, Miller J. Gastro-enteritis, coeliac disease and enamel hypoplasia. Br Dent J 1979; 147: 91–95.
11. Rasmussen P, Espelid I. Coeliac disease and dental malformation. J Dent Child 1980; 47: 190–192.
12. Nikiforuk G, Fraser D. The etiology of enamel hypoplasia: A unifying concept. J Pediatr 1981; 98: 888–893.
13. Gawkrodger DJ, Ferguson A, Barnetson RC. Nutritional status in patients with dermatitis herpetiformis. Am J Clin Nutr 1988; 48: 355–360.
14. Auricchio S, Greco L, Troncone R. What is the true prevalence of coeliac disease? Gastroenterology International 1990; 3: 140–142.
15. Mäki M, Lähdeaho M-L, Hällström O, Viander M, Visakorpi JK. Postpubertal gluten challenge in coeliac disease. Arch Dis Child 1989; 64: 1604–1607.
16. Ferguson A, Blackwell JN, Barnetson RStC. Effects of additional dietary gluten on the small-intestinal mucosa of volunteers and of patients with dermatitis herpetiformis. Scand J Gastroenterol 1987; 22: 543–549.
17. Hallert C, Gotthard R, Norrby K, Walan A. On the prevalence of adult coeliac disease in Sweden. Scand J Gastroenterol 1981; 16: 257–261.
18. Logan RFA, Rifkind EA, Busuttil A, Gilmour HM, Ferguson A. Prevalence and "incidence" of celiac disease in Edinburgh and the Lothian Region of Scotland. Gastroenterology 1986; 90: 334–342.