

# Periodontal therapy in siblings with Papillon–Lefèvre syndrome and tinea capitis: a report of two cases

Schacher B, Baron F, Ludwig B, Valesky E, Noack B, Eickholz P. Periodontal therapy in siblings with Papillon–Lefèvre syndrome and tinea capitis: a report of two cases. J Clin Periodontol 2006; 33: 829–836. doi 10.1111/j.1600-051X.2006.00992.x

#### Abstract

**Objective:** Report of clinical and microbiological periodontal findings before and 6 months after treatment of two siblings with Papillon–Lefèvre syndrome (PLS) and tinea capitis.

**Methods:** Two brothers, RG 3 years and NG 5 years of age, were referred for treatment due to premature mobility of their deciduous teeth. Probing depths (PPD), attachment levels (PAL-V), and furcation involvements were examined clinically. Panoramic radiographs were taken. Subgingival plaque samples within the deepest pocket of each tooth were taken and analysed by real-time polymerase chain reaction (PCR) for *Actinobacillus actinomycetemcomitans (AA)*, *Porphyromonas gingivalis, Tannerella forsythensis, Treponema denticola, Fusobacterium nucleatum*, and *Prevotella intermedia*. One-stage full-mouth scaling and extraction of hopeless teeth were performed under general anaesthesia, followed by systemic amoxicillin and metronidazole for 7 days. Clinical and microbiological analyses were performed 6 months after treatment.

**Results:** Before treatment, both siblings had exhibited PPD of up to 13 mm, Class III furcation defects at four teeth, and marginal suppuration. *AA* was detected in both patients and at all teeth at levels ranging from  $3.0 \times 10^2$  to  $5.1 \times 10^6$ . Both patients exhibited palmar and plantar hyperkeratosis. Seven teeth were extracted from RG, and nine from NG. Six months after treatment, PPD had been reduced to  $\leq 5 \text{ mm}$ . *AA* was not detected in any of the remaining teeth.

**Conclusion:** Even periodontally affected deciduous teeth of PLS patients can be treated successfully. Suppression of *AA* to below detection level seems to be of high significance.

# B. Schacher<sup>1</sup>, F. Baron<sup>1</sup>, B. Ludwig<sup>2</sup>, E. Valesky<sup>3</sup>, B. Noack<sup>4</sup> and P. Eickholz<sup>1</sup>

Departments of <sup>1</sup>Periodontology; <sup>2</sup>Orthodontics, Center for Dental, Oral, and Maxillofacial Medicine; <sup>3</sup>Dermatology, Hospital of the Johann Wolfgang Goethe-University at Frankfurt, 60590 Frankfurt, Germany; <sup>4</sup>Periodontology, Department of Conservative Dentistry, Center for Dental, Oral, and Maxillofacial Medicine, University Hospital, Dresden, Germany

Key words: Actinobacillus actinomycetemcomitans-associated periodontitis; Papillon-Lefèvre syndrome; tinea capitis

Accepted for publication 19 July 2006

Papillon–Lefèvre syndrome (PLS) is characterized by palmar and plantar hyperkeratosis, combined with severe periodontal destruction affecting both the primary and permanent dentitions (Papillon & Lefèvre 1924). The disease is rare (one to four cases per 1 million individuals) and is inherited in an autosomal recessive pattern (Gorlin et al. 1964). PLS may occur in siblings (Preus & Gjermo 1987, Firatli et al. 1996a, Rüdiger et al. 1999, De Vree et al. 2000, Cagli et al. 2005). Microbiological studies of plaque samples from patients with PLS have shown the presence of Gram-negative anaerobic pathogens, often but not always including *Actinobacillus actinomycetemcomitans* (*AA*) (Kressin et al. 1995, Rüdiger et al. 1999, Eickholz et al. 2001, Pacheco et al. 2002).

After localization of a gene for PLS to chromosome11q14–q21 (Fischer et al. 1997, Laass et al. 1997, Hart et al. 1998), it was elucidated that this

syndrome is caused by mutations in the *Cathepsin C (CTSC)* gene (Hart et al. 1999, Toomes et al. 1999). It was shown that certain mutations result in a nearly complete loss of function of the cathepsin C (Toomes et al. 1999). To date, more than 40 mutations of this gene have been identified (Selvaraju et al. 2003, Noack et al. 2004).

Conventional periodontal therapy (oral hygiene instructions, scaling and root planing, periodontal surgery, systemic antibiotics) often failed in PLS patients (Bullon et al. 1993, Hattab et al. 1995, De Vree et al. 2000). Some case reports described the early extraction of all erupted teeth, followed by an edentulous period, to prevent subsequent infection of the non-erupted teeth (Baer & McDonald 1981, Preus 1988, Tinanoff et al. 1995, Wiebe et al. 2001, Ullbro et al. 2005). This concept resulted in successful outcomes, but successful cases have also been described that did not include largescale extractions (Ishikawa et al. 1994, Eickholz et al. 2001, Pacheco et al. 2002, Lux et al. 2005). In these cases, AA had either not been detected before or had been eliminated by treatment.

Other findings beyond skin and periodontal lesions have also been reported for PLS, e.g. immunological changes (Van Dyke et al. 1984, Firatli et al. 1996b, Ghaffar et al. 1999, Lundgren et al. 2005) and increased susceptibility to infection (Haim & Munk 1965, Haneke et al. 1975). Patients with PLS seem to be predisposed to develop pyogenic liver abscesses (Khandpur & Reddy 2001, Almuneef et al. 2003).

Especially in children, tinea capitis is a frequent fungal infection of the skin and hair with involvement of the hair shaft and the pilosebaceous unit. The causative organisms for tinea capitis in Germany are Microsporum (M.) canis (54.8%) and Tircophyton (T.)mentagrophytes (14.7%), followed by T. verrucosum (8.1%), T. violaceum (6.1%), and T. tonsurans (3.8%) (Tietz et al. 1999). The distribution of pathogens shows geographical differences, e.g. in the United States T. tonsurans has been replaced by M. adouinii, and M. canis (Gupta et al. 1999) completely. The clinical pattern of tinea capitis varies: The non-inflammatory type manifests as a subtile clinical infection with one or multiple patches of alopecia, whereas the tinea capitis profunda shows an inflammatory, boggy tender scalp with pustules and with purulent discharge. In addition, the patients may have occipital lymphadenopathy, malaise, and fever (Gupta et al. 1999).

The present report describes the periodontal, dermatological, and genetic findings in two brothers having PLS and tinea capitis. The periodontal treatment of the deciduous dentition with 6-month results and the dermatological treatment are reported.

# Case report

# Patients

Two brothers, RG 3 years and NG 5 years of age, were referred to the Department of Periodontology of the Center for Dental, Oral, and Maxillofacial Medicine at the University Hospital of Frankfurt. The family's dentist had observed premature mobility of the deciduous teeth and asked for additional diagnostics and therapy.

Both the children had palmoplantar hyperkeratosis since the first months of life. Initially, the symptoms started with an erythema of the palms and the soles. Later on, the lesions spread to the back of the hands and feet. Additionally, RG showed psoriasiform plaques on the knees and elbows.

The boys' parents (father, age 42; mother, age 38) are of Eritrean origin, but had been living in Germany for 13 years. Both parents were in generally good health and had not previously had any major or unusual dental or periodontal problems. No unusual dental pathologies were reported in their extended families. The parents are not blood related. Before scientific examination and treatment of the patients, the risks, benefits, and procedures were explained to their parents and informed consent was obtained. The investigation was approved by the Institutional Review Boards for Human Studies of the Universities of Frankfurt and Dresden.

# Clinical and radiographic examination

At the time of the baseline examination, NG and RG presented with complete deciduous dentitions. Generalized supragingival plaque, bleeding as well as suppuration on probing, and class I to III tooth mobility were found in both patients (Fig. 1a and b). Owing to the young age of the patients and the highly inflamed gingival tissues at the initial presentation, a reduced periodontal



Fig. 1. Clinical situation before therapy: (a) NG, (b) RG.



Fig. 2. Panoramic radiographs before therapy: (a) NG, (b) RG.



*Fig. 3.* Tinea capitis in RG: (a) before and (b) 3 months after combined mechanic and antibiotic periodontal therapy: inflammatory erythematosquamous plaques with pustules, purulent discharge, and yellow crusts on the scalp.

examination was performed revealing probing depths (PPD) of 2–9 mm in both and vertical attachment levels (PAL-V) of 2–9 mm in NG.

The panoramic radiographs for NG and RG showing generalized mainly

horizontal bone loss are presented in Fig. 2a and b.

The clinical and radiological findings yielded a diagnosis of periodontitis as a manifestation of PLS for both patients. The children's teeth were professionally cleaned, and they and their mother received comprehensive oral hygiene instructions.

A comprehensive periodontal status, including assessment of PPD and PAL-V at six sites per tooth to the nearest 1 mm (PCPUNC 15, Hu-Friedy, Chicago, IL, USA), was obtained immediately before subgingival debridement under general anaesthesia. Furcation diagnosis (clinical horizontal attachment levels) was performed using a calibrated Nabers probe (PQ2 N, Hu Friedy).

#### Microbiological examination

Immediately before subgingival instrumentation, under general anaesthesia, subgingival plaque samples were obtained from the deepest pocket of each tooth using sterile paper points and analysed for AA, Porphyromonas gingivalis (PG), Tannerella forsythensis (TF), Treponema denticola (TD), Fusobacterium nucleatum (FN), and Prevotella intermedia (PI)using а commercially available real-time polymerase chain reaction (PCR) test (Meridol Paro Diagnostik, Gaba GmbH, Lörrach, Germany).

## Genetic examination

Venous blood samples were taken. After DNA purification (DNA purification kit, Oiagen, Hilden, Germany), all seven exons of the CTSC gene including exon/intron boundary regions were amplified by PCR and directly sequenced as described previously (Noack et al. 2004). Generated sequences (ALFwin Sequence Analyser 2.00 software, Amersham Pharmacia Biotech, Freiburg, Germany) were aligned with published CTSC sequences (GenBank accession no. NT\_08984). DNA sequence variants were confirmed by sequencing at least two independent PCR products.

#### Periodontal treatment

NG's teeth 51 and 85 had been extracted before systematic treatment due to acute inflammation and pain. Systematic treatment commenced for both NG and RG with subgingival scaling using ultrasonic applicators and hand instruments under general anaesthesia. The periodontally hopeless teeth (NG: teeth 55, 54, 64, 74, 72, 71, 81, 82, and 83; RG: teeth 54, 51, 64, 74, 71, 81, and 84) were subsequently removed.

All remaining teeth received a subgingival instillation if a 1% chlorhexidine gel (Chlorhexamed Gel, GlaxoSmith-Kline Consumer Healthcare, Bühl, Germany). The parents were instructed to have the children rinse with 0.12%chlorhexidine solution (GUM Paroex, John O. Butler, Kriftel, Germany) twice daily and to brush their children's teeth with 1% chlorhexidine gel. Both patients received amoxicillin 250 mg and metronidazole 250 mg three times daily for 1 week. Post-operative follow-up examinations with professional tooth cleaning and oral hygiene re-motivation sessions were performed after 1, 2, 3, 6, 10, 14, 18, and 22 weeks. Space maintainers were inserted for NG, 3 weeks post-operatively.

1. 4. 5. 5.

# **Dermatological treatment**

Before periodontal therapy NG and RG were admitted to the department of dermatology for treatment of the hyperkeratotic skin lesions. After combined mechanic and antibiotic periodontal therapy in both siblings, the development of inflammatory erythematosquamous plaques with pustules, purulent discharge, and yellow crusts on the scalp, accompanied by worsening of palmoplantar hyperkeratosis, was observed (Fig. 3).

RG's disease was more pronounced. The clinical diagnosis of tinea capitis was confirmed in RG with the Wood's lamp (filtered ultraviolet light peak of 365 nm). The direct light microscopic examination of RG's hair showed a fungal ectotrix infection, and the subsequent culture (takes 14 days to grow) identified M. canis as the pathogenic organism. In NG, the light microscopic examination and the fungal culture were negative. It is known in the literature that in a substantial number of cases (approximately 50%) the fungal culture can be negative (Silverman & Elewski 1998). Because of the mild manifestation and the negative light microscopic and culture analysis, we decided to treat NG only with an intensive local treatment. The whole scalp was treated with ciclopiroxolamin cream under occlusion twice daily. Furthermore, a selenium sulphide shampoo was used once daily.

RG additionally received itraconazole systemically (5 mg/kg weight) for 4 weeks. No adverse events have been observed. After 1 month of treatment, Table 1. (a) NG's clinical (PPD, probing depths; PAL-V, attachment level; Fu, furcation involvement) and microbiological (log transformed numbers of AA, Actinobacillus actinomycetemcomitans; PG, Porphyromonas gingivalis; TF, Tannerella forsythensis; TD, Treponema denticola; FN, Fusobacterium nucleatum; PI, Prevotella intermedia) parameters before therapy

														_		_		_		_		_							_	
9.7	′x1(	)5	1.(	0x1	0 <sup>5</sup>	3.	6x1	0 <sup>5</sup>	3.	9x1	0 <sup>2</sup>				AA	4.	8x1	0 <sup>5</sup>	5.	4x1	0 <sup>5</sup>	2.9	9x10	0⁴	1.8	3x1	0 <sup>6</sup>	1.6	6x1	0 <sup>5</sup>
1.8	3x10	)⁴	6.3	3x1	07	1.	9x1	0 <sup>4</sup>	1.	1x1	07				PG	1.	3x1	0 <sup>6</sup>	3.	5x1	0 <sup>6</sup>	1.3	3x1(	0 <sup>5</sup>	3.9	łx1	0 <sup>6</sup>	1.2	2x1	0 <sup>6</sup>
4.2	2x10	) <sup>3</sup>	5.:	2x1	0 <sup>6</sup>	4.	4x1	0 <sup>3</sup>	2.	1x1	0 <sup>6</sup>				TF	8.	6x1	0 <sup>5</sup>	3.	2x1	0 <sup>6</sup>	7.8	3x1	0⁴	2.0	)x1	0 <sup>6</sup>	4.	0x1	0 <sup>6</sup>
5.1	x10	)⁴	1.	1x1	07	6.	1x1	0 <sup>3</sup>	1.	5x1	0 <sup>6</sup>				TD	1.	3x1	0 <sup>6</sup>	2.	9x1	0 <sup>6</sup>	2.8	3x1	0 <sup>5</sup>	2.1	ix1	0 <sup>6</sup>	3.	0x1	0 <sup>6</sup>
5.4	x10	)⁴	4.:	2x1	0⁴	1.	7x1	0 <sup>4</sup>	7.	4x1	0 <sup>5</sup>				FN	7.	5x1	0 <sup>3</sup>	2.	9x1	0 <sup>6</sup>	2.	7x1	0 <sup>3</sup>	1.3	3x1	0 <sup>4</sup>	2.4	4x1	0 <sup>6</sup>
5.1	1x10	)⁴	9.4	4x1	0 <sup>6</sup>	2.	3x1	0 <sup>4</sup>	1.	3x1	0 <sup>6</sup>				PI	3.	3x1	0 <sup>6</sup>	1.	9x1	0 <sup>6</sup>	7.0	Dx1	0 <sup>3</sup>	2.4	1x1	0 <sup>6</sup>	1.	9x1	0 <sup>6</sup>
	Ш			Ш											Eu											ш			11	
ш	Τ	1	ш		111																				111		111	11 1		_
10	3	4	6	6	8	4	2	3	3	2	3				PAL-	3	1	2	3	2	2	7	1	4	3	3	4	4	3	7
13	9	8	8	7	7	4	3	4	4	4	3		-		v	4	4	4	4	3	4	6	5	4	8	7	7	1     1.22       1     2.4:0       1     2.4:3       3     3.0:1       1     1.9:1       1     1.9:1       1     1.9:1       1     1.9:1       5     5       5     5       6     6       4     4	10	7
10	3	4	6	6	9	4	2	4	2	2	3					3	2	3	4	2	3	8	2	5	4	3	5	5	3	7
42	П	_	7	ш	-	E		-	5	1			_		PPD	4	"	4	5	1		8	-	5	•	╝	6	8		5
13	8	8	<u> </u>	5	<u>′</u>	э	4	2	2	4	ľ					4	4	-	5	3	1	0	5	5	-	5	Ŭ	1.0     1.0       6     1.2       7     7       5     5       6     8       1     1       4     4       7     7       5     5       6     8       4     4       4     4       4     4       4     4       4     4	9	Ľ
	E E .			54			53			52			51				61			62			63			64			65	
55		_																												
	55					I									1	_				70			70			74		— Г	75	
	85			84			83			82			81				71			72			73	1		74			75	
	85		3	84	4	8	83 8	10	8	82 5	6	4	81 3	4	PPD	6	71	7	5	72 4 11	6	4	73	4	4	74 4 11	4	4	75 6	6
	85		3	84 3 3	4	8 8	83 8 111 7	10 11	8 9	82 5 III 5	6 9	4	81 3 111 2	4	PPD	6	71 4 111 4	7	5 5	72 4 11 2	6 10	4	73 3 1 2	4	4	74 4 11 3	4	1.2 4.0 3.0 2.4 1.9 1 4 5 8 8 6 7 7 5 8 8 6 7 7 4 4 4 4 4 4	75 6 2	6
	85		3 5 3	84 3 3 3	4 4 4	8 8 8	83 8 111 7 5	10 11 10	8 9 8	82 5 111 5 6	6 9 8	4 4 4	81 3 111 2 3	4	PPD	6 6 7	71 4 111 4 5	7 7 6	5 5 4	72 4 11 2 5	6 10 6	4 4 3	73 3 1 2 2	4	435	74 4 11 3 5	4	4 4 4	75 6 2 7	6
	85		3 5 3 6	84 3 3 3 4	4 4 4 4	8 8 8 7	83 8 III 7 5 7	10 11 10 10	8 9 8 9	82 5 111 5 6 9	6 9 8 10	4 4 4 4	81 3 111 2 3 3	4 5 4 5	PPD PAL -V	6 6 7 6	71 4 111 4 5 4	7766	5 5 4	72 4 11 2 5	6 10 6 9	4 4 3	73 3 1 2 2 2	4 4 3	4 3 5 2	74 4 11 3 5	4 4 3	4 4 3	75 6 2 7	6
	85		3 5 3 6	84 3 3 3 4	4 4 4	8 8 8 7	83 8 III 7 5 7	10 11 10 10	8 9 8 9	82 5 111 5 6 9	6 9 8 10	4 4 4 4	81 3 111 2 3 3	4	PPD PAL -V	6 6 7 6	71 4 111 4 5 4	7766	5 5 4	72 4 11 2 5	6 10 9	4 4 3 3	73 3 1 2 2 2	4 4 3	4 3 5 2	74 4 11 3 5 3 111	4 4 3	4 4 3	75 6 2 7 1	6
	85		3 5 3 6	84 3 3 3 4 1	4 4 4	8 8 7	83 8 111 7 5 7	10 11 10 10	8 9 9	82 5 111 5 6 9	6 9 8 10	4 4 4	81 3 111 2 3 3	4	PPD PAL -V Fu	6 6 7	71 4 111 4 5 4	7 7 6	5 5 4	72 4 11 2 5	6 10 6 9	4 4 3 3	73 3 1 2 2 2	4 3 3	4 3 5 2	74 4 11 3 5 3 111	4	4	75 6 2 7 1 1	6
	85		3 5 3 6	84 3 3 3 4 1 1 3x1	4 4 4 0 <sup>4</sup>	8 8 7 7.	83 8 III 7 5 7 9x1	10 11 10 10	8 9 9	82 5 111 5 6 9	6 9 8 10	4 4 4 4	81 3 111 2 3 3 3	4 5 4 5	PPD PAL -V Fu PI	6 6 7 6 7.	71 4 111 4 5 4	7 7 6 6	5 5 4 3.	72 4 11 1 1x1	6 10 9 0 <sup>6</sup>	4 4 3 3	73 3 1 2 2 2 6x1	4 4 3 3	4 3 5 2	74 4 11 3 5 3 111 111 5x1	4 4 3	4 4 1 1.	75 6 2 7 1 1 1	6 4 6 3
	85		3 5 3 6 1. 5.	84 3 3 3 4 1 1 3x1 1x1	4 4 4 4 0 <sup>4</sup> 0 <sup>2</sup>	8 8 7 7. 4.	83 8 III 7 5 7 9x1 9x1	10 11 10 10 0 <sup>5</sup> 0 <sup>4</sup>	8 9 8 9	82 5 111 5 6 9 9	6 9 8 10 0 <sup>5</sup> 0 <sup>5</sup>	4 4 4 4. 2.	81 3 111 2 3 3 3 4x1 0x1	4 5 4 5 0 <sup>5</sup>	PPD PAL -V Fu PI FN	6 6 7 6 7. 2.	71 4 111 4 5 4 1x1 6x1	7 6 6 0 <sup>5</sup> 0 <sup>4</sup>	5 5 4 4 3. 1.	72 4 1 2 5 1 1 1x1 8x1	6 10 6 9 0 <sup>6</sup> 0 <sup>3</sup>	4 4 3 3 2.	73 3 1 2 2 2 6x1 6x1	4 4 3 3	4 3 5 2 1. 3.	74 4 3 5 3 III 5x1 5x1	4 4 3	4 4 3 1. 7.	75 6 2 7 1 1 7x1 4x1	6 4 6 3 0 <sup>7</sup>
	85		3 5 3 6 1. 5. 1.	84 3 3 3 4 1 1x1 2x1	4 4 4 0 <sup>4</sup> 0 <sup>2</sup> 0 <sup>5</sup>	8 8 7 7. 4. 1.	83 8 III 7 5 7 7 9x1 9x1 9x1 4x1	10 11 10 10 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>6</sup>	8 9 8 9 8 3	82 5 111 5 6 9 9 2x1 2x1 2x1	6 9 8 10 0 <sup>5</sup> 0 <sup>5</sup>	4 4 4 4. 2.	81 3 111 2 3 3 3 4x1 0x1 7x1	4 5 4 5 0 <sup>5</sup> 0 <sup>5</sup>	PPD PAL -V Fu PI FN TD	6 6 7 6 7. 2. 4.	71 4 111 4 5 4 4 1x1 6x1 7x1	7 6 6 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>5</sup>	5 5 4 4 3. 1. 5.	72 4 11 2 5 1 1 1x1 8x1 5x1	6 10 6 9 0 <sup>6</sup> 0 <sup>3</sup> 0 <sup>6</sup>	4 4 3 3 2. 1. 8.	73 3 1 2 2 2 6x1 6x1 9x1	4 4 3 3 0 <sup>6</sup> 0 <sup>6</sup>	4 3 5 2 1. 3. 3.	74 4 3 5 3 III 5x1 5x1 0x1	4 4 4 3	4 4 3 1. 7. 7.	75 6 2 7 1 1 1 1 7x1 7x1	6 4 6 3 0 <sup>7</sup> 0 <sup>5</sup> 0 <sup>6</sup>
	85		3 5 3 6 1. 5. 1. 1.	84 3 3 3 4 1 1x1 2x1 2x1	4 4 4 4 0 <sup>4</sup> 0 <sup>2</sup> 0 <sup>5</sup> 0 <sup>5</sup>	8 8 7 7. 4. 1. 8.	83 8 111 7 5 7 7 9x1 9x1 4x1 1x1	10 11 10 10 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>6</sup> 0 <sup>5</sup>	8 9 8 9 8 3. 1, 7	82 5 111 5 6 9 9 2x1 2x1 2x1 2x1 5x1	6 9 8 10 0 <sup>5</sup> 0 <sup>5</sup> 0 <sup>5</sup>	4 4 4 4. 2. 1. 2.	81 3 111 2 3 3 3 4x1 0x1 7x1 6x1		PPD PAL -V Fu PI FN TD TF	6 6 7 6 7. 2. 4. 4.	71 4 111 4 5 4 1x1 6x1 7x1 7x1 4x1	7 7 6 6 6 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>5</sup> 0 <sup>4</sup>	5 5 4 4 3. 1. 5. 2.	72 4 1 2 5 1 1x1 8x1 5x1 5x1	6 10 6 9 0 <sup>6</sup> 0 <sup>8</sup> 0 <sup>6</sup>	4 4 3 2. 1. 8. 3.	73 3 1 2 2 2 6x1 6x1 6x1 9x1 5x1	4 4 3 3 0 <sup>6</sup> 0 <sup>6</sup> 0 <sup>6</sup>	4 3 5 2 1. 3. 3. 2.	74 4 3 5 3 111 5x1 5x1 5x1 7x1	4 4 3 10 <sup>7</sup> 10 <sup>6</sup>	4 4 3 1. 7. 7. 6.	75 6 2 7 1 1 1 7x1 4x1 7x1 4x1 7x1	
	85		3 5 3 6 1. 5. 1. 1. 2.	84 3 3 3 3 4 1 1x1 2x1 2x1 0x1	$ \begin{array}{c} 4 \\ 4 \\ 4 \\ 4 \\ 0^{4} \\ 0^{2} \\ 0^{5} $	8 8 7 7. 4. 1. 8. 4.	83 8 111 7 5 7 7 9x1 9x1 9x1 9x1 4x1 1x1 6x1	10 11 10 10 $0^{5}$ $0^{4}$ $0^{6}$ $0^{5}$ $0^{6}$	8 9 8 9 8 3 1. 7. 1.	82 5 111 5 6 9 9 2x1 2x1 2x1 5x1 5x1	6 9 8 10 0 <sup>5</sup> 0 <sup>5</sup> 0 <sup>6</sup> 0 <sup>6</sup>	4 4 4 4. 2. 1. 2. 3.	81 3 111 2 3 3 3 4x1 0x1 7x1 6x1 1x1	4 5 4 5 $0^5$ $0^5$ $0^5$ $0^5$	PPD PAL -V Fu PI FN TD TF PG	6 6 7 6 7. 2. 4. 4. 1.	71 4 5 4 1x1 6x1 7x1 4x1 3x1		5 5 4 4 3. 1. 5. 2. 1.	72 4 1 2 5 1 1 1x1 8x1 5x1 5x1 5x1 4x1	6 10 6 9 0 <sup>6</sup> 0 <sup>3</sup> 0 <sup>6</sup> 0 <sup>7</sup>	4 4 3 2. 1. 8. 3.	73 3 1 2 2 2 2 2 2 2 6x1 6x1 6x1 9x1 5x1 2x1	4 4 3 3 0 <sup>6</sup> 0 <sup>6</sup> 0 <sup>6</sup> 0 <sup>7</sup>	4 3 5 2 1. 3. 3. 2. 4.	74 4 3 5 3 111 5x1 5x1 5x1 7x1 5x1	4 4 3 10 <sup>7</sup> 10 <sup>6</sup> 10 <sup>6</sup>	4 4 3 1. 7. 7. 6. 7.	75 6 2 7 1 1 1 4x1 7x1 7x1 0x1 2x1	

the light microscopy and consecutive culture were negative.

The follow-up assessment showed minimal clinical signs of tinea capitis in RG and none in NG. The therapy with local ciclopiroxolamin cream was conducted in both siblings for a further 4 weeks. The palmoplantar keratoderma was treated by intensive topical therapy using 10-20% urea in an ointment and cream base twice daily as the basis treatment. The hyperkeratosis decreased almost completely.

### Examination of the parents

Both parents were also clinically examined, panoramic radiographs were obtained, subgingival plaque samples were taken at the deepest site of each quadrant, and analysed using the RNA probe test (IAI Pado-Test 4.5<sup>®</sup>, Institut für angewandte Immunologie, Zuchwil, Switzerland). Finally, samples of venous blood were obtained.

### Results

### Clinical and microbiological parameters

The detailed clinical and microbiological results for NG and RG are presented in Table 1 and Table 2: before treatment, markedly increased PPD (up to 13 mm) were found for nearly all teeth, with the findings more pronounced for NG than for RG (Table 1). Six months after treatment, PPDs were generally reduced to 2-4 mm with one site in either sibling exhibiting PPD of 5 mm (Table 2).

Before treatment, AA at levels of  $3.0 \times 10^2 - 5.1 \times 10^6$  had been detected in NG and RG (Table 1). Six months after treatment, AA was not detectable in either patient (Table 2).

*Table 1.* (continued) (b) RG's clinical (PPD, probing depths; PAL-V, attachment level; Fu, furcation involvement) and microbiological (log transformed numbers of AA, *Actinobacillus actinomycetem-comitans*; PG, *Porphyromonas gingivalis*; TF, *Tannerella forsythensis*; TD, *Treponema denticola*; FN, *Fusobacterium nucleatum*; PI, *Prevotella intermedia*) parameters before therapy.

Γ	5.0	)x1	)²	3.6	5x1	0 <sup>4</sup>	4	.0x1	0 <sup>5</sup>	1.	7x1	0 <sup>4</sup>	2.0	)x10	)5	AA	8.:	2x1	0⁴	9	8x10	<b>)</b> ⁴	2.	5x1	0 <sup>5</sup>	1.	5x1	0 <sup>4</sup>	4       1.2:         6       2.7:         5       1.5:         5       5.0:         3       4.0:         6       7.5:         11       1         8       2         7       2         7       4         7       4         6       3         7       6         3       4	2x1(	) <sup>5</sup>
F	7.3	3x1(	)²	6.2	2x1	0 <sup>6</sup>	4	.7x1	0 <sup>4</sup>	1.	3x1	0 <sup>5</sup>	1.8	3x10	)5	PG	8.	5x1	0 <sup>5</sup>	5	.0x10	) <sup>3</sup>	6.	6x1	0⁴	2.	5x1	$0^4$ $0^6$ $0^5$ $0^5$ $0^6$ III 8 7 8 7 6 6	2.	7x1(	) <sup>5</sup>
F	<2.	5x1	0 <sup>2</sup>	1.3	2x1	0 <sup>6</sup>	9	.1x1	0 <sup>4</sup>	9.	7x1	0 <sup>3</sup>	4.2	2x10	)⁴	TF	1.4	4x1	0 <sup>5</sup>	3	.7x1(	D⁴	3.	7x1	0 <sup>4</sup>	6.	1x1	0 <sup>4</sup> 0 <sup>6</sup> 0 <sup>5</sup> 0 <sup>7</sup> 10 <sup>8</sup> 111 8 7 7 8 7	1.	5x1(	כ5
Γ	2.9	9x1	0 <sup>2</sup>	1.1	7x1	0 <sup>4</sup>	6	.2x1	0 <sup>4</sup>	3.	2x1	0 <sup>3</sup>	6.8	3x10	) <sup>3</sup>	тD	3.	9x1	0 <sup>4</sup>	4	.8x1(	)²	7.	2x1	0 <sup>2</sup>	3.	3x1	0 <sup>5</sup>	5.	0x1(	0⁴
	<2.	5x1	0 <sup>2</sup>	1.	5x1	0 <sup>5</sup>	3	.2x1	0 <sup>5</sup>	1.	4x1	0 <sup>4</sup>	8.1	x10	) <sup>3</sup>	FN	1.4	4x1	0 <sup>4</sup>	1.3x10⁴		2.6x10⁴			3.1x10 <sup>3</sup>			4.	0x1(	24	
F	<2.	5x1	0 <sup>2</sup>	4.6	3x1	06	1.0x1		0 <sup>5</sup>	7.	7x1	7x10⁴		3x10	)⁴	Ы	6.	1x1	0 <sup>5</sup>	3.9x10 <sup>3</sup>		4.4x10 <sup>5</sup>			3.0x10 <sup>6</sup>			7.5x1		כ5	
Γ					111																				111		L				
				Ξ												Fu						_				Ш		Ш			
Γ	4	3	3	6	8	7	2	1	2	3	3	5	8	7	8	PAL-	6	6	6	5	5	3	2	3	3	4	5	8	2	2	1
	2	2	5	7	7	7	4	2	2	4	4	5	7	5	5	v	5	4	5	2	1	2	1	2	1	8	5	7	2	1	1
┢	-	2	-		4	7		2			3	6		5	£		5	5	6	6	6		2	4	1	5	5			2	2
	5	Ū	э	0	Ť	'	4	-	4	4	1	Ľ	Ļ		5	PPD	5	-	0	0	1	Ļ	l°.	-	Ļ	5	11	0	Ľ		3
	4	3	5	7	7	7	6	4	4	5	5	6	8	5	6		6	5	6	4	3	4	4	5	3	8	4	7	4	3	4
F		55			54		53					52		51			61			62			63			64					
_															_																
		85			84			83			82			81				71			72			73			74	_		75	
			_					1 2						3	- I			E	6		2			2					12	2	4
	3	3	4	3	4	5	3	₽°	3	3	3	3	4	-	5	000	7	5	۱°	17	<u> </u>	3	3	-	4	5	4	6	ľ	_	
	3	3	4	3 5	4	5 5	3 5	5	3	3 4	3	3	4	3	5	PPD	7 6	5 III 6	6	3	1	3	3 2	3	4	5 5	4    5	6 5	4	3	3
	3 3 2	3 2 2	4 4 2	3 5 3	4    4 5	5 5 5	3 5 2	5	3 4 2	3 4 2	3             	3 4 2	4 4 3	3 3	5 4 5	PPD	7 6 8	5     6 7	6	4 3	 3 2	3	3 2 2	3	4 5 2	5 5 4	4    5 5	6 5 6	4	3	3
	3	3 2 2	4 4 2	3 5 3	4    4 5	5 5 5	3 5 2	5	3	3 4 2	3           	3	4	33	5 4 5	PPD PAL-	7 6 8	5     6 7	6	4	 3 2	3	3 2 2	3	4	5 5 4	4    5 5	6 5 6	4	3	3
	3 2 1	3 2 2 1	4 4 2 2	3 5 3 4	4    4 5 4	5 5 4	3 5 2 4	5 1 4	3 4 2 3	3 4 2 3	3               	3 4 2 4	4 4 3 4	333	5 4 5 4	PPD PAL- V	7 6 8 7	5 III 6 7 6	6 8 6	4 3 4 2	 3 2 2	3 2 2 2	3 2 2 0	3	4	5 5 4 4	4    5 5 6	6 5 6 7	4 2 1	3 2 1	3 3 1
	3 3 2 1	3 2 2 1	4 4 2 2	3 5 3 4	4 11 4 5 4 111	5 5 4	3 5 2 4	5 1 4	3 4 2 3	3 4 2 3	3               	3 4 2 4	4 3 4	333	5 4 5 4	PPD PAL- V	7 6 8 7	5 III 6 7 6	6 8 6	4 4 2	 3 2 2	3 2 2	3 2 2 0	3	4	5 4 4	4    5 5 6 	6 5 6 7	4 2 1	3 2 1	3
	3 3 2 1	3 2 2 1	4 4 2 2	3 5 3 4	4    4 5    4 	554	3 5 2 4	5	3 4 2 3	3 4 2 3	3 1 4 2 4	3	4 3 4	333	5 4 5 4	PPD PAL- V Fu	7 6 8 7	5     6 7 6	6 8 6	4 2	1 3 2 2	3 2 2	3 2 2 0	3	4	5 4 4	4    5 5 6 	6 5 6 7	4 2 1	3 2 1	3
	3 3 2 1	3 2 2 1 0x11	4 4 2 2	3 5 3 4 1.	4    4 5    4         1x1	5 5 4 0 <sup>6</sup>	3 5 2 4 1	5 1 4	3 4 2 3	3 4 2 3 3	3   4   2   4	3 4 2 4	4 3 4 6.4	3 3 3	5 4 5 4	PPD PAL- V Fu PI	7 6 8 7 5.	5 111 6 7 6 3x1	6 8 6	4 2 1	 3 2 2	3 2 2	3 2 0	3 1 3	4 5 2 4	5 4 4 2.	4    5 5    6         2x1	6 5 6 7	4 2 1 2.	3 2 1 3x1	3 3 1
	3 3 2 1 1.	3 2 2 1 0x10 1x10	4 2 2 0 <sup>5</sup>	3 5 3 4 1. 2.	4    4 5    4     1x1 5x1	5 5 4 0 <sup>6</sup> 0 <sup>3</sup>	3 5 2 4 1 <2	.8x1	3 4 2 3 0 <sup>4</sup> 10 <sup>2</sup>	3 4 2 3 3 <2	3 1 4 2 4 4	3 4 2 4	4 4 3 4 6.4	3 3 3 1x1( 2x1(	5 4 5 4	PPD PAL- V Fu PI FN	7 6 8 7 5.	5 111 6 7 6 3x1 5x1	6 8 0 <sup>5</sup> 0 <sup>4</sup>	4 2 1 3	1 3 2 2 .4x1	3 2 2 0 <sup>6</sup> 0 <sup>2</sup>	3 2 2 0 1 <1	3 1 3	4 5 2 4	5 4 4 2. 7.	4    5 5    6         2x1 3x1	6 5 6 7 0 <sup>5</sup> 0 <sup>3</sup>	4 2 1 2. 4.	3 2 1 3x11 5x11	3 3 1
	3 3 2 1 1. 1. 2.	3 2 2 1 0x10 1x10 8x10	4 4 2 2 0 <sup>5</sup> 0 <sup>4</sup>	3 5 3 4 1. 2. 1.	4    4 5 4     1x1 5x1 1x1	5 5 4 0 <sup>6</sup> 0 <sup>3</sup> 0 <sup>5</sup>	3 5 2 4 1 2 5	.8x1 2.5x <sup>2</sup>	3 4 2 3 0 <sup>4</sup> 10 <sup>2</sup> 0 <sup>2</sup>	3 4 2 3 3 3 <2 <2	3 1 4 2 4 4 .3x1 2.5x <sup>4</sup>	3 4 2 4	4 4 3 4 6.4 1.2 6.7	3 3 3 3 1x10 2x10 7x10	$5 \\ 4 \\ 5 \\ 4 \\ 5^{5} \\ 3^{3} \\ 3^{3}$	PPD PAL- V Fu FN TD	7 6 8 7 5. 1. 2.	5 111 6 7 6 3x1 5x1 4x1	6 8 6 0 <sup>5</sup> 0 <sup>4</sup>	4 2 1 3 <2	1 3 2 2 2 .4x11 .9x11 2.5x1	3 2 2 0 <sup>6</sup> 0 <sup>2</sup> 0 <sup>2</sup>	3 2 0 1 <1 6	3 1 .0x1 .8x1	4 5 2 4	5 4 4 2. 7. 1.	4    5 5 (6)     2x1 3x1 8x1	6 5 6 7 0 <sup>5</sup> 0 <sup>3</sup> 0 <sup>4</sup>	4 2 1 2. 4. 3.	3 2 1 3x10 5x10 4x10	3 3 1 0 <sup>4</sup> 0 <sup>2</sup>
	3 3 2 1 1. 1. 2. 1.	3 2 2 1 0x10 1x10 8x10 4x10	4 2 2 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>3</sup>	3 5 3 4 1. 2. 1.	4    4 5    4     1x1 5x1 1x1	5 5 4 0 <sup>6</sup> 0 <sup>5</sup> 0 <sup>6</sup>	3 5 2 4 1 <2 5 8	.8x1 2.5x <sup>-</sup> .4x1 .1x1	3 4 2 3 3 0 <sup>4</sup> 10 <sup>2</sup> 0 <sup>2</sup> 0 <sup>2</sup>	3 4 2 3 3 2 2 2	3 1 4 2 4 4 .3x1 2.5x <sup>4</sup> .5x <sup>4</sup>	3 4 2 4 0 <sup>5</sup> 10 <sup>2</sup> 0 <sup>3</sup>	4 4 3 4 6.4 1.2 6.7 1.9	3 3 3 3 1x10 2x10 7x10	5 4 5 4 5 4 0 <sup>5</sup> 0 <sup>3</sup> 0 <sup>3</sup> 0 <sup>3</sup>	PPD PAL- V Fu PI FN TD TF	7 6 8 7 5. 1. 2. 7.	3 111 6 7 6 3x1 5x1 4x1 4x1	6 8 6 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>4</sup>	4 2 1 3 <2 2	. 1 3 2 2 .4x11 .9x11 .9x11	3 2 2 2 0 <sup>6</sup> 0 <sup>2</sup> 0 <sup>2</sup> 0 <sup>4</sup>	3 2 2 0 1 <1 6 <1	3 1 .2x1 .0x1 .0x1	4 5 2 4 4	5 4 4 2. 7. 1. 3.	4    5 5 ( 6     2x1 3x1 8x1 8x1	6 5 6 7 7 0 <sup>5</sup> 0 <sup>3</sup> 0 <sup>4</sup> 0 <sup>5</sup>	4 2 1 2. 4. 3. 9.	3 2 1 3x10 5x10 4x10 4x10	3 3 1 $0^4$ $0^3$ $0^2$ $0^3$
	3 3 2 1 1. 1. 1. 2. 1. 4.0	3 2 2 1 0x10 1x10 8x10 6x10	4 4 2 2 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>4</sup> 0 <sup>4</sup>	3 5 3 4 1. 2. 1. 1. 4.	4 11 4 5 4 111 111 111 111 111	5 5 4 0 <sup>6</sup> 0 <sup>5</sup> 0 <sup>6</sup> 0 <sup>6</sup>	3 5 2 4 1 <2 5 8 1	.8x1 .1x1 .5x1	3 4 2 3 0 <sup>4</sup> 10 <sup>2</sup> 0 <sup>2</sup> 0 <sup>2</sup> 0 <sup>4</sup>	3 4 2 3 3 <2 <2 2 1	3 1 4 2 4 4 3x1 2.5x <sup>4</sup> .5x <sup>4</sup> .5x <sup>4</sup>	3 4 2 4 4 0 <sup>5</sup> 10 <sup>2</sup> 0 <sup>3</sup> 0 <sup>3</sup>	4 4 3 4 6.4 1.2 6.7 1.9 1.1	3 3 3 3 1 2 1 2 1 2 2 1 2 2 1 1 2 2 1 1 2 2 1 1 2 2 1 1 2 2 1 1 2 2 2 1 2 2 2 1 2 2 2 1 2 2 1 2	$5$ 4 5 4 $5^{5}$ $3^{3}$ $3^{3}$ $3^{5}$	PPD PAL- V Fu PI FN TD TF PG	7 6 8 7 5. 1. 2. 7. 8.	3 III 6 7 6 3x1 5x1 4x1 4x1 4x1	6 8 6 0 <sup>5</sup> 0 <sup>4</sup> 0 <sup>4</sup> 0 <sup>5</sup>	4 2 1 3 <2 5	1 3 2 2 2 .4x11 2.5x1 .9x11 .9x11 .9x11	3 3 2 2 $0^{6}$ $0^{2}$ $0^{2}$ $0^{4}$ $0^{4}$	3 2 2 0 1 <1 6 <1 1	3 1 3 .2x1 .0x1 .0x1 .0x1 .8x1	4 5 2 4 4 0 <sup>5</sup> 0 <sup>2</sup> 0 <sup>2</sup> 0 <sup>2</sup>	5 4 4 2. 7. 1. 3. 1.	4    5 5    6     2x1 3x1 8x1 8x1 1x1	6 5 6 7 7 0 <sup>5</sup> 0 <sup>3</sup> 0 <sup>4</sup> 0 <sup>5</sup> 0 <sup>6</sup>	4 2 1 2. 4. 3. 9. 2.	3 2 1 3x10 5x10 4x10 4x10 1x10	3 3 1 $0^4$ $0^2$ $0^2$ $0^3$ $0^4$

The parents' clinical and radiologic findings were each consistent with a generalized moderate chronic periodontitis. AA was not detected microbiologically in sub-gingival plaque samples of either parent.

## **Genetic parameters**

Sequence analysis of the *CTSC* gene identified a functionally relevant homozygote missense mutation in NG and RG in exon 5 (c.755A>T, p.Q252L) first described by Toomes et al. (1999). Both parents were heterozygous carriers of the c.755A>T mutation in the *CTSC* gene.

### Discussion

The present report documents the diagnostic measures and partial preservation of teeth in two cases of generalized periodontitis in the deciduous dentition of two brothers with PLS. To the best of our knowledge, this is the first report of subgingival instrumentation and partial preservation of deciduous teeth in treatment of PLS periodontitis. Both patients were referred with severe periodontal inflammation including bleeding and suppuration on probing, premature mobility of deciduous teeth, attachment loss, and furcation involvement (class III). NG had acute symptoms with pain and periodontal abscesses at two teeth. Both siblings also exhibited palmoplantar hyperkeratoses. The clinical diagnosis of PLS was confirmed by genetic analysis showing a corresponding mutation of the CTSC gene that was found to be homozygotic in the children and heterozygotic in their parents.

Detailed microbiological examination revealed subgingival presence of AA at all teeth before treatment, confirming the observation that this periodontal pathogen plays a key role in the aetiology of periodontitis in PLS (Kleinfelder et al. 1996, De Vree et al. 2000, Eickholz et al. 2001, Wara-aswapati et al. 2001, Lux et al. 2005). Owing to the fact that AA could not be detected in either of the parents, the source of the AA-infection remains in question. On the one hand, it cannot be ruled out totally that the parents harbour AA subgingivally because microbiological sampling was performed only at the four teeth with most pronounced PPD. On the other hand, the source of infection may be located in other relatives who were not available for examination. However, on this issue we can only speculate.

Combined mechanical and antibiotic periodontal therapy resulted in drastic clinical improvements in both patients and suppressed AA to below the quite low detection limit of real-time PCR  $(10^2)$ . This primary result is in agreement with the results of other researchers (Rüdiger et al. 1999, Eickholz et al. 2001, Pacheco et al. 2002). Presumably, the positive treatment outcome may partially be attributed to the fact that mechanical therapy, including the necessary extractions, followed the principle of full-mouth disinfection (Quirynen et al. 1995). Amoxicillin and metronidazole, administered concurrently, are reliable and effective in the treatment of AA-associated periodontitis (Van Winkelhoff et al. 1989), which was confirmed in the present case.

Thus, an overall favourable development may be expected. This presupposes, however, that the young patients are closely followed, with their parents intricately involved, as the children are not yet able to ensure adequate oral hygiene on their own.

Wiebe et al. (2001) reported a successful long-term therapeutic outcome in the case of a young boy with PLS who had undergone periodontal treatment since the age of 4, despite mediocre oral hygiene. In that case, the periodontal infection could only be eliminated by extracting all deciduous teeth and a subsequent edentulous phase after metronidazole monotherapy had failed. Subsequent microbiological analysis of the permanent dentition showed no indication of AA. Extraction of all periodontally involved teeth causing an edentulous phase seems to be a reliable strategy to eliminate AA. However, this

*Table 2.* (a) NG's clinical (PPD, probing depths; PAL-V, attachment level; Fu, furcation involvement) and microbiological (log transformed numbers of AA, *Actinobacillus actinomy-cetemcomitans*; PG, *Porphyromonas gingivalis*; TF, *Tannerella forsythensis*; TD, *Treponema denticola*; FN, *Fusobacterium nucleatum*; PI, *Prevotella intermedia*) parameters 5 months after therapy

					0			0		Γ			AA		0			0			0						0	
				1.	9x1	0 <sup>2</sup>	7.5	5x1	0 <sup>1</sup>				PG	9.	5x1	0 <sup>1</sup>	1.0	0x1	0 <sup>2</sup>	9.	8x1	0 <sup>1</sup>				1.8	8x1	02
				1.	6x1	0 <sup>2</sup>	3.	0x1	0 <sup>2</sup>				TF	1.0	0x1	0 <sup>2</sup>	1.	4x1	0 <sup>2</sup>		0					1.7	7x1	0 <sup>3</sup>
					0		2.:	2x1	0 <sup>2</sup>				TD	2.0	6x1	0 <sup>1</sup>	9.8x10 <sup>1</sup>		2.0x10 <sup>1</sup>		0 <sup>1</sup>				2.1x10		0 <sup>3</sup>	
				5.0x10 <sup>1</sup>			2.9x10 <sup>1</sup>					FN	3.:	2x1	0 <sup>3</sup>	8.:	2x1	0 <sup>3</sup>	5.	0x1	0 <sup>4</sup>				2.4	4x1	0⁴	
				1.	6x1	0 <sup>1</sup>	2.0	6x1	0 <sup>2</sup>				Ы		0			0			0						0	
				1																								
Γ			Γ	1	0	0	1	1	2				PAL-	3	2	3	2	2	3	3	1	2				1	1	3
<b> </b>  -	_		┢	2		1	1		1		-		V	2	2	3	2	1	2	2	2	3				2	4	2
				2	2	2	2	2	2				1	2	2	3	3	2	3	4	2	3				3	2	3
$\vdash$	+		┢	3	-	3	3	-	2		-	-	PPD	3	 	3	4	1	3	3	2	3				3	3	2
	<u> </u>			┝	- <u>-</u>			52			51	<u> </u>		-	61			62			63			64	L		65	<u> </u>
	5		•	L				52			-		J							L						L		
8	15	84	ļ	[	83			82	-		41		]	Γ	31			72			73			74			75	
		2 2	2							3	2	3		3	2	3				3	2	2			ļ	3	2	3
T-	-	2 2	3	F				-		3	3	3	PPD	3	3	2		┡		2	2	2		-		3	2	3
		2 2	2							0	0	0		0	0	0				3	1	1			<u> </u>	2	2	2
	-	3 2	2			-				0	<u> </u>	0	V	0	-	0		1	╞	2	-	3		-		1	-	2
<u> </u>	L	2	I		<u> </u>				L		0		-	-	0	1	╞	I		┢	2		-		I	-	•	
													Fu										-			-		
		0								9.	5x1	0 <sup>2</sup>	РІ	4.	1x1	10 <sup>2</sup>					0						0	
		1.24	104							2.	2x1	0 <sup>2</sup>	FN	3.	0x1	10 <sup>2</sup>	Γ			1.	4x1	<b>0</b> ⁴				2.	9x1	0 <sup>3</sup>
		1.27	10											_			-			1								
		0		╞						4.	2x1	0 <sup>2</sup>	TD	2.	5x1	10 <sup>3</sup>				1.	3x1	101				1.	4x1	03
F		0								4. 5.	2x1 1x1	0 <sup>2</sup> 0 <sup>2</sup>	TD TF	2. 1.	5x1 5x1	10 <sup>3</sup>	$\left  \right $			1.	3x1 0	10 <sup>1</sup>				1. 1.	4x1 8x1	0 <sup>3</sup> 0 <sup>3</sup>
		0 0 6.9x	10 <sup>1</sup>							4. 5. 2.	2x1 1x1 1x1	0 <sup>2</sup> 0 <sup>2</sup> 0 <sup>2</sup>	TD TF PG	2. 1. 3.	5x1 5x1 1x1	10 <sup>3</sup> 10 <sup>3</sup> 10 <sup>2</sup>				1. 1.	3x1 0 5x1	10 <sup>1</sup>				1. 1. 8.	4x1 8x1 5x1	0 <sup>3</sup> 0 <sup>3</sup> 0 <sup>1</sup>

is a quite drastic measure possibly having psychological consequences for the children and likely to cause orthodontic problems due to the lack of space-maintaining function of the deciduous teeth (Lux et al. 2005).

As AA was no longer found on the deciduous teeth of either of the two boys treated, it may be assumed that the eruption of the first permanent teeth, which is currently in progress in NG, may occur in a healthy periodontal environment. The risk of AA reinfection of the children is considered to be low, as the pathogen was not found in either of the parents. The orthodontic consequences due to the unavoidable loss of the most severely involved teeth were taken care of by insertion of space maintainers.

Ahuja et al. (2005) described the case of a 10-year-old PLS patient: after scaling and root planing, combined with systemic administration of amoxicillin and metronidazole, failed to improve the periodontal situation, flap surgeries with a second antibiotic treatment was successful. Despite systemic amoxicillin and metronidazole, it may be hypothesized that in this case, AA could not be eliminated by non-surgical therapy. Persistence of AA despite repeated mechanical therapy combined with amoxicillin and metronidazole had been described before by Rüdiger et al. (1999). Finally, renewed antibiotics combined with surgical therapy with its additional potential to remove AA from the tissue were successful. However, Ahuja et al. (2005) performed microbiological sampling neither before nor after therapy. Thus, we cannot be sure of the reason behind the initial failure and the final success of therapy.

On the one hand, the systemic use of retinoids has been reported to be effective in patients with PLS (Lee et al. 2005, Sethuraman et al. 2005). On the other, retinoid treatment may cause skeletal abnormalities in children, slender long bones and premature epiphyseal closure as well as mucocutaneous dryness, abnormal liver function tests, and elevated triglycerides (Lee et al. 2005). Some authors reported that systemic retinoids not only improve skin lesions but also gingival inflammation by an unknown mechanism (Lee et al. 2005, Sethuraman et al. 2005). Others demonstrated that the level of periodontal infection and the severity of skin affection do not correlate at all (Ullbro et al. 2003). Because of the mild to moderate manifestation of the palmoplantar keratoderma and the young age of the patients, we initially decided to use only local dermatological treatment in both of them.

Tinea capitis, which had aggravated in both children after combined mechanical and antibiotic treatment, is to be considered a separate disease. It can only be speculated that children with PLS associated with abnormal functional or quantitative neutrophils in 50% may be more susceptible to dermatomycoses. Furthermore, the medication with systemic antibiotics (amoxicillin and metronidazole) may have supported the growth of the dermatophytes by changing bacterial and fungal skin flora quantitatively as well as qualitatively (Brook 2000).

The dental treatment provider of young PLS patients should make sure to keep an eye on the children's general health and to ensure that they see a paediatrician or appropriate specialist as required.

To the best of our knowledge, this is the first report of (i) PLS with a manifestation of tinea capitis and (ii) of subgingival instrumentation and at least short-term partial preservation of deciduous teeth in the treatment of PLS periodontitis.

# Acknowledgement

This study was kindly supported by GABA GmbH, Lörrach, Germany.

3

3

3

0

Table 2. (continued) (b) RG's clinical (PPD, probing depths; PAL-V, attachment level; Fu, furcation involvement) and microbiological (log transformed numbers of AA, Actinobacillus actinomycetemcomitans; PG, Porphyromonas gingivalis; TF, Tannerella forsythensis; TD, Treponema denticola; FN, Fusobacterium nucleatum; PI, Prevotella intermedia) parameters 5 months after therapy.

		0	0		1	0	0	0		0
0		0	0			0	0	0		
1.9x10 <sup>°</sup>		2.7x10 <sup>2</sup>	1.0x10 <sup>-</sup>		PG	1.6x10 <sup>-</sup>	8.2x10	8.9x10		1.9x10-
4.7x10 <sup>2</sup>		1.7x10 <sup>2</sup>	9.2x10'		TF	0	0	1.6x10 <sup>2</sup>		0
1.8x10 <sup>5</sup>		2.2x10 <sup>2</sup>	3.7x10 <sup>2</sup>		TD	6.9x10°	3.5x10⁴	0		8.8x10 <sup>3</sup>
2.0x10 <sup>6</sup>		9.0x10 <sup>3</sup>	1.1x10 <sup>5</sup>		FN	1.5x10⁵	4.3x10⁴	1.0x10⁴		5.7x10 <sup>5</sup>
0		0	0		PI	1.8x10 <sup>2</sup>	0	2.6x10 <sup>1</sup>		0
					Fu					
0 1 0		0 0 1	1 2 4		PAL-	4 3 2	4 1 3	2 1 1		1 0 1
1 0 1		1 0 1	2 1 2		V	3 2 2	2 1 1	0 1 0		2 1 1
3 2 2		2 2 3	3 2 4		1	4 3 2	5 1 4	3 2 3		3 2 3
3 2 3		3 2 3	4 3 4		PPD	4 4 4	4 3 3	2 3 2		4 3 3
55	54	53	52	51	1	61	62	63	64	65
					-					
85	84	83	82	81	]	71	72	73	74	75
85 2 2 3	84	83 2 2 3	82 2 2 2	81		71	72 2 2 2 1	73 3 2 2	74	75 3 3 3
85 2 2 3 4 2 3	84	83 2 2 3 2 2 3	82 2 2 2 3 2 2	81	- PPD	71	72 2 2 2 1 3 2 3	73 322 322	74	75 3 3 3 3 2 3
85 2 2 3 4 2 3 0 0 1	84	83 2 2 3 2 2 3 0 0 1	82 2 2 2 1 3 2 2 2 2 2 2 2	81	PPD	71	72 2 2 2 3 2 3 2 2 1	73 3 2 2 3 2 1 0 1	74	75 3 3 3 3 2 1 1
85 2 2 3 4 2 3 0 0 1 2 0 1	84	83       2     2       3       2     2       3       0     0       1	82       2     2       3     2       2     2       2     2       2     2       2     2       2     2	81	PPD PAL -V	71	$\begin{array}{c cccc} 72 \\ 2 & 2 \\ 3 & 2 \\ 2 & 1 \\ 3 & 2 \\ 2 & 1 \\ 1 & 0 \\ 1 & 0 \\ 1 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0 \\ 0$	73       3     2       3     2       1     0       1     0	74	75 3 3 3 3 2 1 1 -
85         2         2         3           4         2         3         3         3           0         0         1         1         2         0         1	84	83       2     2       3       2     2       3       0     0       1	82       2     2       3     2       2     2       2     2       2     2       2     2       3     2	81	PPD PAL -V		72       2     2       3     2       3     2       1     0	73       3     2       3     2       3     2       1     0       1     0	74	75       3     3       3     2       1     1       0     0
85           2         2         3           4         2         3           0         0         1           2         0         1	84	83       2     2       3       2     2       3       0     0       1	82       2     2       3     2       2     2       2     2       2     2       2     2       3     3	81	PPD PAL -V Fu	71	$ \begin{array}{c ccccccccccccccccccccccccccccccccccc$	73       3     2       3     2       3     2       1     0       1     0		75 3 3 3 3 2 1 1 0 0 0
85           2         2         3           4         2         3           0         0         1           2         0         1	84	83       2     2       3     3       2     2       3     3       0     0       1     1       0     1       0     0	82           2         2         2           3         2         2           2         2         2           2         2         2           2         2         3	81	PPD PAL -V Fu PI		72       2     2       3     2       3     2       1     0       1     0	73       3     2       3     2       1     0       1     0		75       3     3       3     2       1     1       0     0
85           2         2         3           4         2         3           0         0         1           2         0         1           2         0         1           0         4.5x10 <sup>4</sup> 1	84	83 2 2 3 2 2 3 0 0 1 0 1 1 0 1 1 0 3.6x10 <sup>4</sup>	82 2 2 2 2 3 2 2 2 2 2 2 2 2 2 2 3 0 1.7x10 <sup>6</sup>	81	PPD PAL -V Fu FN		72 2 2 2 3 2 3 2 2 1 1 0 1 0 1.3x10 <sup>6</sup>	73       3     2       2     2       3     2       1     0       1     0       0     0       2.5x10 <sup>5</sup>		75 3 3 2 1 1 0 0 0 2.0x10 <sup>6</sup>
85           2         2         3           4         2         3           0         0         1           2         0         1           2         0         1           0         4.5x10 <sup>4</sup> 1.8x10 <sup>3</sup>	84	83           2         2         3           2         2         3           0         0         1           0         1         1           0         3.6x10 <sup>4</sup> 0	82           2         2         2           1         2         2           2         2         2           2         2         2           2         2         2           3         -         -           2         2         2           3         -         -           2         2         2           3         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -         -         -           -	81	PPD PAL -V Fu PI FN TD		72 2 2 2 2 3 2 3 2 2 1 1 0 1 1 0 1 	73       3     2       2     2       3     2       1     0       1     0       0     2.5x10 <sup>5</sup> 5.3x10 <sup>1</sup>		75 3 2 3 1 1 0 0 0 0 2.0x10 <sup>6</sup> 1.0x10 <sup>6</sup>
85           2         2         3           4         2         3           0         0         1           2         0         1           2         0         1           4         2         3           0         0         1           2         0         1           0         4.5x10 <sup>4</sup> 1.8x10 <sup>3</sup> 0         0         1	84	83           2         2         3           2         2         3           0         0         1           0         1         1           0         3.6x10 <sup>4</sup> 0           0         0         0	82           2         2         2           1         2         2           2         2         2           2         2         2           2         2         2           2         2         2           1.7x10 <sup>6</sup> 0           0         0	81	PPD PAL -V Fu FN TD TF		72 2 2 2 2 3 2 3 2 2 1 1 0 1 1 0 1 1.3x10 <sup>6</sup> 5.5x10 <sup>2</sup> 0	73       3     2       2     2       3     2       1     0       1     0       0     2.5x10 <sup>5</sup> 5.3x10 <sup>1</sup> 4.8x10 <sup>1</sup>		75 3 2 3 1 1 0 0 0 0 2.0x10 <sup>6</sup> 1.0x10 <sup>6</sup> 0
85           2         2         3           4         2         3           0         0         1           2         0         1           2         0         1           0         4.5x10 <sup>4</sup> 1.8x10 <sup>3</sup> 0         1.4x10 <sup>2</sup> 1		83 2 2 3 2 2 3 0 0 1 1 1 0 1 1 0 3.6x10 <sup>4</sup> 0 0 7.9x10 <sup>1</sup>	82 2 2 2 1 3 2 2 2 2 2 2 2 2 2 2 2 2 2 2 2 2 3 0 1.7x10 <sup>6</sup> 0 0 1.2x10 <sup>2</sup>	81	PPD PAL PAL PI FN TD TF PG		72       2     2       3     2       3     2       3     2       1     0       1     0       1.3x10 <sup>6</sup> 5.5x10 <sup>2</sup> 0       1.5x10 <sup>2</sup>	73       3     2       2     2       3     2       2     2       1     0       1     0       0     2.5x10 <sup>5</sup> 5.3x10 <sup>1</sup> 4.8x10 <sup>1</sup> 9.8x10 <sup>1</sup>		75 3 2 3 1 1 0 0 0 0 2.0x10 <sup>6</sup> 1.0x10 <sup>5</sup> 0 5.0x10 <sup>2</sup>
85           2         2         3           4         2         3           0         0         1           2         0         1           2         0         1           0         4.5x10 <sup>4</sup> 3           0         1.8x10 <sup>3</sup> 0           1.4x10 <sup>2</sup> 0         1		83           2         2         3           2         2         3           0         0         1           0         1         1           0         1         1           0         3.6x10 <sup>4</sup> 0           0         0         7.9x10 <sup>1</sup> 0         0         1	82       2     2       3     2       2     2       2     2       2     2       2     2       3     2       2     2       2     2       3     1.7x10 <sup>6</sup> 0     0       1.2x10 <sup>2</sup> 0	81	PPD PAL -V Fu FN TD TF PG AA		$\begin{array}{c ccccccccccccccccccccccccccccccccccc$	73           3         2         2           3         2         2           1         0         1           1         0         0           2.5x10 <sup>5</sup> 5.3x10 <sup>1</sup> 4.8x10 <sup>1</sup> 9.8x10 <sup>1</sup> 0         0		75 3 2 3 1 1 0 0 0 0 2.0x10 <sup>6</sup> 1.0x10 <sup>6</sup> 0 5.0x10 <sup>7</sup> 0

### References

- Ahuja, V., Shin, R. H., Mudgil, A., Nanda, V. & Schoor, R. (2005) Papillon-Lefèvre syndrome: a successful outcome. Journal of Periodontology 76, 1996-2001.
- Almuneef, M., Khenaizan, S. A., Ajaji, S. A. & Al-Anazi, A. (2003) Pyogenic liver abscess and Papillon-Lefèvre syndrome: not a rare association. Pediatrics 111, 85-88.
- Baer, P. N. & McDonald, R. E. (1981) Suggested mode of periodontal therapy for patients with Papillon-Lefèvre syndrome. Periodontal Case Reports 1, 10.
- Brook, I. (2000) The effects of amoxicillin therapy on skin flora in infants. *Pediatric* Dermatology 17, 360-363.
- Bullon, P., Pascual, A., Fernandez-Novoa, M. C., Borobio, M. V., Muniain, M. A. & Camacho, F. (1993) Late onset Papillon-Lefèvre syndrome? A chromosomic, neutrophil function and microbiological study.

Journal of Clinical Periodontology 20, 662-667.

- Cagli, N. A., Hakki, S. S., Dursun, R., Tov, H., Gokalp, A., Ryu, O. H., Hart, P. S. & Hart, T. C. (2005) Clinical, genetic, and biochemical findings in two siblings with Papillon-Lefèvre syndrome. Journal of Periodontology 76, 2322-2329.
- De Vree, H., Steenackers, K. & de Boever, J. A. (2000) Periodontal treatment of rapid progressive periodontitis in 2 siblings with Papillon-Lefèvre syndrome: 15-years follow-up. Journal of Clinical Periodontology 27, 354-360.
- Eickholz, P., Kugel, B., Pohl, S., Näher, H. & Staehle, H. J. (2001) Combined mechanical and antibiotic periodontal therapy in a case of Papillon-Lefèvre syndrome. Journal of Periodontology 72, 542-549.
- Firatli, E., Gurel, N., Efeoglu, A. & Badur, S. (1996a) Clinical and immunological find-

ings in 2 siblings with Papillon-Lefèvre syndrome. Journal of Periodontology 67, 1210-1215.

- Firatli, E., Gurel, N., Efeoglu, A. & Badur, S. (1996b) Papillon-Lefèvre syndrome. Analysis of peripheral blood lymphocyte subsets. Journal of Clinical Periodontology 23, 823-825.
- Fischer, J., Blanchet-Bardon, C., Prud'homme, J. F., Pavek, S., Steijlen, P. M., Dubertret, L. & Weissenbach, J. (1997) Mapping of Papillon-Levèvre syndrome to the chromosome 11q14 region. European Journal of Human Genetics 5, 156-160.
- Ghaffar, K. A., Zahran, F. M., Fahmy, H. M. & Brown, R. S. (1999) Papillon-Lefèvre syndrome. Neutrophil function in 15 cases from 4 families in Egypt. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology and Endodontics 88, 320-325.
- Gorlin, R. J., Sedano, H. & Anderson, V. E. (1964) The syndrome of palmar-plantar hyperkeratosis and premature periodontal destruction of the teeth. The Journal of Pediatrics 65, 895-906.
- Gupta, A. K., Hofstader, S. L., Adam, P. & Summerbell, R. C. (1999) Tinea capitis: an overview with emphasis on management. Pediatric Dermatology 16, 171-189.
- Haim, S. & Munk, J. (1965) Keratosis palmoplantaris congenita, with peridontosis, arachnodactyly and a peculiar deformity of the terminal phalanges. British Journal of Dermatology 77, 42-54.
- Haneke, E. (1975) The Papillon-Lefèvre syndrome: keratosis palmoplantaris with periodontopathy. Human Genetics 51, 1-35.
- Hart, T. C., Bowden, D. W., Ghaffar, K. A., Wang, W., Cutler, C. W., Cebeci, I., Efeoglu, A. & Firatli, E. (1998) Sublocalization of the Papillon-Lefèvre syndrome locus on 11g14q21. American Journal of Medical Genetics 79, 134-139.
- Hart, T. C., Hart, P. S., Bowden, D. W., Michalec, M. D., Callison, S. A., Walker, S. J., Zhang, Y. & Firatli, E. (1999) Mutation of the cathepsin C gene are responsible for Papillon-Lefèvre syndrome. Journal of Medical Genetics 36, 881-887.
- Hattab, F. N., Rawashdeh, M. R., Yassin, O. M., Al-Momani, A. S. & Al-Ubosi, M. M. (1995) Papillon-Lefèvre syndrome: a review of the literature and a report of 4 cases. Journal of Periodontology 66, 413-420.
- Ishikawa, I., Umeda, M. & Laosrisin, N. (1994) Clinical, bacteriological, and immunological examination and the treatment process of two Papillon-Lefèvre syndrome patients. Journal of Periodontology 65, 364-371.
- Khandpur, S. & Reddy, B. S. (2001) Papillon-Lefèvre syndrome with pyogenic hepatic abscess: a rare association. Pediatric Dermatology 18, 45-47.
- Kleinfelder, J. W., Topoll, H. H., Preus, H. R., Müller, R. F., Lange, D. E. & Böcker, W. (1996) Microbiological and immunohistological findings in a patient with Papillon-Lefèvre syndrome. Journal of Clinical Periodontology 23, 1032-1038.

- Kressin, S., Herforth, A., Preis, S., Wahn, V. & Lenard, H. G. (1995) Papillon–Lefèvre syndrome–successful treatment with a combination of retinoid and concurrent systemic periodontal therapy: case report. *Quintessence International* 26, 795–803.
- Laass, M. W., Hennies, H. C., Preis, S., Stevens, H. P., Jung, M., Leigh, I. M., Wienker, T. F. & Reis, A. (1997) Localization of a gene for Papillon–Lefèvre syndrome to chromosome 11q14-q21 by homozygosity mapping. *Human Genetics* 101, 376–382.
- Lee, M. R., Wong, L. C. & Fischer, G. O. (2005) Papillon–Lefèvre syndrome treated with acitretin. *The Australasian Journal of Dermatology* 46, 199–201.
- Lundgren, T., Parhar, R. S., Renvert, S. & Tatakis, D. N. (2005) Impaired cytotoxicity in Papillon–Lefèvre syndrome. *Journal of Dental Research* 84, 414–417.
- Lux, C. J., Kugel, B., Komposch, G., Pohl, S. & Eickholz, P. (2005) Orthodontic treatment in a patient with Papillon–Lefèvre syndrome. *Journal of Periodontology* **76**, 149–157.
- Noack, B., Görgens, H., Hoffmann, T., Fanghänel, J., Kocher, T., Eickholz, P. & Schackert, H. K. (2004) Novel mutations in the Cathepsin C Gene in patients with prepubertal aggressive periodontitis and Papillon– Lefèvre syndrome. *Journal of Dental Research* 83, 368–370.
- Pacheco, J. J., Coelho, C., Salazar, F., Contreras, A., Slots, J. & Velazco, C. H. (2002) Treatment of Papillon–Lefèvre syndrome periodontitis. *Journal of Clinical Periodontology* 29, 370–374.
- Papillon, M. M. & Lefèvre, P. (1924) Deux cas de kératodermie palmaire et plantaire symétrique familiale (maladie de Meleda) chez le frère et la sœur. Coexistence dans les deus cas d'alterations dentaires graves. Bulletin de la Société Française de Dermatologie et de Syphiligraphie **31**, 82–87.
- Preus, H. R. (1988) Treatment of rapidly destructive periodontitis in Papillon–Lefèvre syndrome. *Journal of Clinical Periodontology* 15, 639–643.

# **Clinical Relevance**

*Scientific rationale for study*: PLS is a condition that, if untreated, will lead unevitably to total tooth loss before the age of 30. For therapy in affected deciduous dentitions, extraction of all teeth with an eden-

- Preus, H. R. & Gjermo, P. (1987) Clinical management of prepubertal periodontitis in 2 siblings with Papillon–Lefèvre syndrome. *Journal* of Clinical Periodontology 14, 156–160.
- Quirynen, M., Bollen, C. M. L., Vandekerckhove, B. N. A., Dekeyser, C., Papaioannou, W. & Eyssen, H. (1995) Full- versus partialmouth disinfection in the treatment of periodontal infections: short-term clinical and microbiological observations. *Journal of Dental Research* 74, 1459–1467.
- Rüdiger, S., Petersilka, G. & Flemming, T. F. (1999) Combined systemic and local antimicrobiol therapy of periodontal disease in Papillon–Lefèvre syndrome. *Journal of Clinical Periodontology* 26, 847–854.
- Selvaraju, V., Markandaya, M., Prasad, P., Sathyan, P., Sethuraman, G., Srivastava, S. C., Thakker, N. & Kumar, A. (2003) Mutation analysis of the cathepsin C gene in Indian families with Papillon–Lefèvre syndrome. *BMC Medical Genetics* 4, 5.
- Sethuraman, G., Malhotra, A. K., Khaitan, B. K. & Sharma, V. K. (2005) Effectiveness of isotretinoin in Papillon–Lefevre syndrome. *Pediatric Dermatology* 22, 378–379.
- Silverman, R. A. & Elewski, B. E. (1998) Pediatric mycoses. In: *Cutaneous Fungal Infections*, 2nd edition, pp. 261–285. Malden: Blackwell Science.
- Tinanoff, N., Tempro, P. & Maderazo, E. G. (1995) Dental treatment of Papillon–Lefèvre syndrome: 15 year follow-up. *Journal of Clinical Periodontology* 22, 609–612.
- Tietz, H. J., Czaika, V., Ulbricht, H. M. & Sterry, W. (1999) Tinea capitis in Germany. A survey in 1998. *Mycoses* 42, 73–76.
- Toomes, C., James, J., Wood, A. J., Wu, C. L., McCormick, D., Lench, N., Hewitt, C., Moynihan, L., Roberts, E., Woods, C. G., Markham, A., Wong, M., Widmer, R., Ghaffar, K. A., Pemberton, M., Hussein, I. R., Temtamy, S. A., Davies, R., Read, A. P., Sloan, P., Dixon, M. J. & Thakker, N. S. (1999) Loss-of-function mutations in the cathepsin C gene result in periodontal disease and palmoplantar keratosis. *Nature Genetics* 23, 421–424.

tulous period is still recommended. This invasive concept has several negative orthodontic and psychological consequences.

*Principal findings*: By extraction of hopeless teeth as well as combined mechanical and systemic antibiotic

- Ullbro, C., Brown, A. & Twetman, S. (2005) Preventive periodontal regimen in Papillon– Lefèvre syndrome (in process citation). *Pediatric Dental Journal* **27**, 226–232.
- Ullbro, C., Crossner, C. G., Nederfors, T., Alfadley, A. & Thestrup-Pedersen, K. (2003) Dermatologic and oral findings in a cohort of 47 patients with Papillon–Lefevre syndrome. *Journal of the American Academy of Dermatology* **48**, 345–351.
- Van Dyke, T. E., Taubman, M. A., Ebersole, J. L., Haffajee, A. D., Socransky, S. S., Smith, D. J. & Genco, R. J. (1984) The Papillon– Lefèvre syndrome: neutrophil dysfunction with severe periodontal disease. *Clinical Immunology and Immunopathology* 31, 419–429.
- Van Winkelhoff, A. J., Rodenburg, J. P., Goené, R. J., Abbas, E. G., Winkel, E. G. & de Graaff, J. (1989) Metronidazole plus amoxicillin in the treatment of *Actinobacillus actinomycetemcomitans*-associated periodontitis. *Journal of Clinical Periodontology* 16, 128–131.
- Wara-aswapati, N., Lertsirivorakul, J., Nagasawa, T., Kawashima, Y. & Ishikawa, I. (2001) Papillon–Lefèvre syndrome: serum immunoglobulin G (IgG) subclass antibody response to periodontapathic bacteria. A case report. *Journal of Periodontology* 72, 1747–1753.
- Wiebe, C. B., Häkkinen, L., Putkins, E. E., Walsh, P. & Larjava, H. S. (2001) Successful periodontal maintenance of a case with Papillon–Lefèvre syndrome: 12-year follow-up and review of the literature. *Journal of Periodontology* **72**, 824–830.

Address:

B. Schacher Department of Periodontology Johann Wolfgang Hospital Goethe-University at Frankfurt Theodor-Stern-Kai 7 60590 Frankfurt Germany E-mail: schacher@em.uni-frankfurt.de

therapy, clinical parameters were improved and AA was suppressed below detection limit.

*Practical implication*: PLS periodontitis in deciduous dentitions may be treated successfully without extracting all teeth.

This document is a scanned copy of a printed document. No warranty is given about the accuracy of the copy. Users should refer to the original published version of the material.