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SHORT COMMUNICATION

Neurofibromatosis 1 and dental caries

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Abstract A total of 110 patients with neurofibromatosis 1 (NF1) were evaluated for their dental health. Appropriate cohorts from national Finnish databases were used as reference. The results showed that NF1 patients presented lower rate of caries compared to controls in age groups under 35 years. The differences between NF1 patients and the reference population diminished by age. In conclusion, (1) NF1 per se does not predispose to caries; and (2) even if NF1 had an adverse effect on dental health, poor outcome can be counteracted with good personal dental care supported by well organized primary health care. The results of the present study are important to report since a common anecdotal perception is that the rate of caries may be higher in NF1 compared to reference population.

Keywords Bone · Caries · Dental health · DMFT · Neurofibromatosis · Teeth

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Introduction

Neurofibromatosis 1 (NF1) is a neurocutaneous skeletal disorder with variable phenotypic expression and an incidence of 1:3000 worldwide [1, 2]. The diagnosis of NF1 is based on criteria outlined by the National Institute of Health (USA) Consensus Development Conference in 1987 [3]. Osseous dysplasias in neurofibromatosis may include reduced bone mineral density, scoliosis, and congenital pseudarthrosis; NF1-related craniofacial findings include, e.g., macrocephaly and sphenoid wing dysplasia. The facial skeletal malformations are results of both genetic and local factors, such as tumor growth [1, 4]. Oral involvement is reported to occur in 3.4–92% of adult patients and in about 40% of children with NF1 [5]. Impacted, displaced, supranumerary, or missing teeth, particularly in the mandible and overgrowth of the alveolar ridge are recognized oral manifestations in NF1. Furthermore, specific lytic bone lesions of the jaws, periapical cemental dysplasia, have been reported to be common in women with NF1 [6]. The rate of dental caries has remained a topic of controversy. This is the first large-scale clinical study focusing on the topic of dental caries rate in NF1.

Patients and methods

A total of 110 patients with NF1, 54 female patients aged 3–68 years and 56 male patients aged 8–73 years, were included in the present study. The study was approved by the Ethics Committee of Southwest Finland Hospital District, Turku, Finland, and the examination was performed with appropriate written consents. All patients fulfilled the diagnostic criteria for NF1 [3]. The patients recruited to the study were among the patients attending the

Table 1 The proportion of subjects with present or past caries or fillings (dmft/DMFT>0) in NF1 patients and in reference subjects (ref) of 861,700 Finnish children under 18 years of age [8]

Age category in years		dmft/DMFT>0		Number of patients
NF1	Ref	NF1 (%)	Ref (%)	NF1
3–4	3	0	16	5
9–11	9	18	60	11
12–14	12	21	62	14
15–17	15	20	76	10
18–20	18	10	84	10

NF1 clinic at the Department of Dermatology, Turku University Central Hospital, as well as among members of the Finnish NF patient organization. This resulted in participation of patients from all parts of Finland. There was no bias to recruit patients suffering from mandibular involvement or any particular dental problem.

The patients were examined in the Department of Oral Diseases, Turku University Central Hospital. The craniofacial and all dental examinations were carried out by the same clinician (V.V.) between years 2005 and 2008. The craniofacial status was based on physical inspection, cephalographs, and orthopantomographs supplemented with periapical radiographs when necessary. In clinical examination, the number of children's teeth was calculated, and the patients were examined for dental decay. Caries was searched using a sharp explorer, fiber optic transillumination, and mouth mirror. Caries was recorded as decayed (d;D), missing (m;M), and/or filled (f;F) teeth (t;T), resulting in dmft/DMFT index (d;D+m;M+f;F/t;T), according to the WHO (1979) criteria [7]. Lower case refers to deciduous teeth and upper case to permanent teeth. The proportion of subjects with index values >0 (dmft/DMFT>0) represents the proportion of subjects with present or past caries experience, i.e., a cumulative prevalence of caries. Two national Finnish studies were available for reference. Report on 861,700 persons covered most children under 18 years of age within the public health care between years 1970 and 2000 [8]. A random sample of 8,028 persons represented adults over 30 years of age [9].

Results and discussion

The present study includes results of clinical examination of 110 patients with NF1. To our knowledge, this is the largest cohort of NF1 patients whose dental examination has been reported to date. The study visits were free, and the traveling expenses were compensated. Thus, the study attracted NF1 patients from all parts of the country, and there was no bias to recruit patients with or without dental problems. However, the current study may have attracted patients who show interest in taking good care of their oral hygiene. The reference populations consisted of two large Finnish national cohorts.

A total of 50 NF1 patients under 20 years of age were included in the study. A general notion was that dental health in individuals with NF1 was good or even better than in the Finnish population in general as estimated by dmft/DMFT index. In the reference group, the cumulative prevalence value (DMFT>0) increased from 60% to 84% between age cohorts 9 and 18 years, while the respective prevalence values varied between 0% and 21%, in the NF1 patients (Table 1). This was somewhat a surprising finding since selected factors and individual cases might suggest the opposite. Specifically, delays in the development of motoric skills are common in NF1, and the children may experience difficulties in brushing their teeth properly. Perhaps the recognition of these problems has prompted both the dentists and parents of the NF1 children to pay more attention to this issue and assist the children with oral hygiene.

Ten NF1 patients were between the ages of 21–29 years, and 10 NF1 patients were 35–44 years old. These groups did not have reference groups. The DMFT index of NF1 individuals ranged from 5.7 to 6.2.

In age categories 30≥65 years, the results present males and females separately (Table 2). A general notion was that the mean numbers of DMFT increased gradually by age category. In the category 30–34 years, the mean values were clearly lower in the NF1 patients than the reference group. In the two oldest age categories, there were no differences between the NF1 patients and the reference group in relation to the numbers of DMF-teeth. The present

Table 2 DMFT in relation to age categories in NF1 patients and the adult reference group [9] in Finland

Dental caries	30–34years				45–54years				65+years			
	Males		Females		Males		Females		Males		Females	
	NF1	Ref	NF1	Ref	NF1	Ref	NF1	Ref	NF1	Ref	NF1	Ref
DMFT	1.8	15.1	9.0	15.1	24.2	23.1	20.2	24.5	29.2	26.4	26.2	27.3
Number of subjects	5	1015	8	1133	9	787	8	837	5	345	5	459

results do not, however, give any indication of increase in dental decay in the NF1 patients.

A previous study has reported increased level of caries in NF1 patients in Canada [10]. In this previous study, questionnaires regarding dental caries were sent to families that included at least one individual with NF1. Siblings with NF1 reported significantly more dental caries than siblings without NF1 in these families. This is in striking contrast to the current study in which the patients were examined by a professional dentist under clinical conditions: The diagnosis of caries and recognition of composite (white) fillings require professional set up with adequate dental instruments such as a sharp explorer, fiber optic transillumination, and mouth mirror.

Conclusions

In conclusion, clinical examination of 110 NF1 patients and comparison with a reference population show that increased caries is not a typical feature in NF1.

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Conflict of interest The authors declare that they have no conflict of interest.

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