

Prevalence of dental caries in children with neurofibromatosis 1

Erica S. Tsang · Patricia Birch · Jan M. Friedman · Douglas Johnston · Tracy Tucker · Linlea Armstrong

Received: 7 October 2009 / Accepted: 1 December 2009 / Published online: 5 January 2010
© Springer-Verlag 2009

Keywords Neurofibromatosis 1 · Dental caries · Oral health · Teeth

Neurofibromatosis 1 (NF1), which affects one in 3,500 individuals, is a progressive autosomal dominant disorder characterized by café-au-lait spots, neurofibromas, optic gliomas, and osseous abnormalities [1]. The oral health of NF1 patients has not yet been fully investigated, but a recent careful study in Finland reported a lower incidence of caries on dental examination in NF1 patients under 35 years of age, as compared to age group controls [2]. This result conflicted with an earlier family study done by our group showing increased self-reported caries in family members with NF1 when compared to their unaffected siblings [3].

As part of a separate sibling-controlled study designed to characterize further the bone health of NF1-affected children, we collected data by dental examination on the oral health status of 18 patients with NF1 and their unaffected siblings. One child with NF1 and one unaffected sibling in each family were studied in an attempt to minimize the effects of factors unrelated to NF1, such as background genetics, diet, and oral hygiene habits, that may

impact dental health. Our participant families came from British Columbia, and the majority of participants were from middle class families who were well informed about the importance of dental hygiene. Eighteen children with NF1 and one unaffected sibling of each child, 6–20 years of age, were enrolled. All but one affected child represented a new mutation. The mean ages of affected children and their unaffected siblings were 13.9 ± 3.3 years (range 6.5–18.9) and 14.9 ± 3.9 years (range 6.7–20.6), respectively. The affected child's age ranged from 4.1 years younger to 4.4 years older than the unaffected control sib. Of the 19 girls enrolled, eight were affected, and 11 were unaffected.

All children underwent dental examinations, during which the numbers of past and present caries were recorded using a decayed, missing, or filled teeth (dmft/DMFT) index. The dmft/DMFT index, also employed by the Finnish group, refers to the number of decayed (d;D), missing (m;M), or filled (f;F) deciduous (lower case letters) or permanent (upper case letters) teeth [4].

Results of the dental examinations are summarized in Table 1. We found a similar prevalence of carries in the affected subjects (8/18) and their unaffected sibs (5/18). Our data do not support the lower prevalence of caries in NF1 patients recently reported by Visnapuu et al. [2] or the higher prevalence we observed in a previous sib-controlled questionnaire study of 37 NF1 patients [3].

In addition to attempting to replicate the simple binary analysis performed by the Finnish group, we compared the number of carious teeth per subject in our paired samples. Neither analysis showed any difference in the prevalence of dental caries in affected and unaffected sibling pairs ($p=0.22$, McNemar's test; $p=0.76$, Wilcoxon signed ranks test).

A direct comparison of our data to those from the Finnish group for NF1 patients is presented in Table 2. We

E. S. Tsang (✉) · P. Birch · J. M. Friedman · T. Tucker · L. Armstrong

Department of Medical Genetics, University of British Columbia,
Box 153, 4500 Oak Street,
Vancouver, BC V6H 3N1, Canada
e-mail: ertsang@interchange.ubc.ca

D. Johnston
Department of Pediatric Dentistry, Children's Hospital,
4480 Oak Street,
Vancouver, BC V6H 3N1, Canada

Table 1 The number of children with past and present caries in among probands with NF1 and their unaffected siblings from British Columbia, Canada

	Sibling with NF1	Unaffected sibling
Number of children with dmft or DMFT >0	8	5
Number of children with dmft or DMFT=0	10	13

Fisher's exact probability (two-tailed)=0.49

selected the 45 Finnish children whose age range (9–20 years) was most similar to our 18 NF1 probands. The prevalence of dental caries was lower among the Finnish patients, and this difference is of borderline statistical significance ($p=0.052$) despite the small number of subjects included in the comparison.

We do not know why our findings differ from those of Visnapuu et al. using similar methods in patients with a similar condition. If this reflects environmental, dietary, or genetic differences between the population studies, it suggests that, although good dental care is key for quality oral health, this alone may not be sufficient in NF1-affected individuals. Further investigation of caries incidence in a greater number of people with NF1 is required to better characterize this risk.

Table 2 The number of children with past and present caries using data for children ages 9–20 from Visnapuu et al. (Finland) in comparison to probands in this study

	Finnish study	British Columbia study
Number of children with dmft or DMFT >0	8	8
Number of children with dmft or DMFT=0	37	10

Fisher's exact probability (two-tailed)=0.052

Acknowledgments This work was supported by the British Columbia Neurofibromatosis Foundation and the British Columbia Children's Hospital Telethon award. We would like to extend special thanks to all study participants.

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

References

1. Gutmann DH, Alysworth A, Carey JC, Korf B, Marks J, Pyeritz RE, Rubenstein A, Viskochil D (1997) The diagnostic evaluation and multidisciplinary management of neurofibromatosis 1 and neurofibromatosis 2. *JAMA* 278:51–57
2. Visnapuu V, Pienihäkkinen K, Peltonen S, Happonen RP, Peltonen J (2009) Neurofibromatosis 1 and dental caries. *Clin Oral Invest* doi:10.1007/s00784-009-0341-x
3. Tucker T, Birch P, Savoy DM, Friedman JM (2007) Increased dental caries in people with neurofibromatosis 1. *Clin Genet* 72:524–527
4. WHO (1979) A guide to oral health. Epidemiological investigations, Geneva

Answer of the authors

On NF1 and dental caries

Vivian Visnapuu^{a,c}, Kaisu Pienihäkkinen^d, Sirkku Peltonen^b, Risto-Pekka Happonen^{c,d}, and Juha Peltonen^{a,b}

^aDepartment of Cell Biology and Anatomy, University of Turku, Kiinamylynkatu 10, FIN 20520 Turku, Finland

^bDepartment of Dermatology, University of Turku, P.O. BOX 52, FIN 20521 Turku, Finland

^cDepartment of Oral Diseases, Turku University Central Hospital, Lemminkäisenkatu 2, FIN 20520 Turku, Finland

^dInstitute of Dentistry, University of Turku, Lemminkäisenkatu 2, FIN 20520 Turku, Finland

The Letter to the Editor by Tsang et al. was prompted in part by our recent article Neurofibromatosis 1 and dental caries (Visnapuu et al., *Clin Oral Invest*). In this report, we showed that increased caries is not a characteristic feature of NF1. The conclusions were drawn from the clinical examination by a dentist of 110 patients with NF1. Fifty of these were under 20 years of age. The findings of the young NF1 patients were compared to a large population database which represents the whole Finnish population under 18 years. Our results were contradictory to findings reported earlier (Tucker et al. 2007). This discrepancy has prompted Tsang et al. to collect data by dental examination of 18 patients with NF1 and their unaffected siblings. The statistics of the study by Tsang et al. also favor the new understanding that caries is not more common in NF1 than in controls. Thus, we do not see the new results from Canada being contradictory to our results. The noted differences in the dental health between the Finnish and Canadian patients can be explained by the special attention to the preventive dental care in Finland. All young age groups in our study have been subjected to well-organized preventive dental care, which has been effective since the late 1970s. Specific attention has been paid to the children who have chronic diseases or developmental delays. The discussion on dental caries in NF1 is extremely valuable not only for the sake of the patients with NF1 but also because of other rare diseases potentially affecting the ability of the patients to take care of their oral hygiene. We are pleased that our work has attracted attention and stimulated re-evaluation of this issue.