Dentinogenesis imperfecta in adults with osteogenesis imperfecta

H GJØRUP[12] (*), JD HALD[3], MH SCHMIDT[1], T HARSLØF[3], L FOLKESTAD[4], BL LANGDAHL[3], D HAUBEK[1]

[1] Department of Dentistry, Health, Aarhus University, Denmark, [2] Section of Oral Health in Rare Diseases, Department of Maxillofacial Surgery, and [3] Department of Medical Endocrinology, Aarhus University Hospital, Denmark, [4] Department of Endocrinology, Odense University Hospital, Denmark

Email: hangjo@rm.dk

Introduction

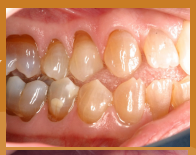
Osteogenesis imperfecta (OI) is a rare inherited disease characterized by fragile bone. In severe cases, bone fractures are frequent incidents. Basically, the disease is a bone dysplasia with abnormalities in the formation of Collagen I. Mainly, OI type I is characterized by a quantitative collagen defect. OI type III and IV are characterized by qualitative a collagen defect. Collagen I is also a substantial part of the dentine of the teeth. Thus, dentinogenesis imperfecta (DI) is likely to be present in patients with OI[1,2]. Teeth with DI are characterized by a greyish or brownish discoloration of the dental crowns and by obliterated pulp chambers in addition to other radiological aberrations.

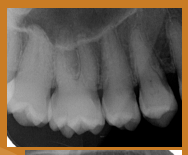
The aim of the study was to describe the prevalence and the clinical variation of DI in adults with OI and to describe dental radiological findings adjunctive to DI.

Materials and Methods

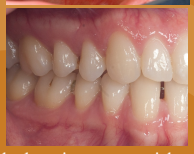
The study population was 73 Danish adults (39 females and 34 males) with OI. Mean age was 47.7 yrs (SD 15.1; range 22-80 yrs). The OI-diagnosis was classified according to Sillence 1979 (OI type I, III, or IV) and the diagnosis was genetically verified. By cultured dermal fibroblasts, the collagen of the patient was characterized as normal or quantitatively or qualitatively deficient.
The study material included clinical photos of the dentition, a panoramic radiograph, periapical radiographs of the teeth, and bitewings.

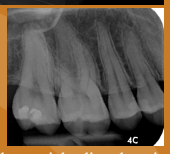
OI type IV with DI





OI type I without DI





For the individual patient, a greyish or brownish discoloration of the dental crowns was assessed as indicative of DI.
Radiological signs indicative of DI were defined as
- Obliterated pulp chambers
- Cervical constrictions
- Short roots

The radiographs were assessed according to these criteria. Adjunctive radiographic signs of dental pathologies or anomalies were noticed.

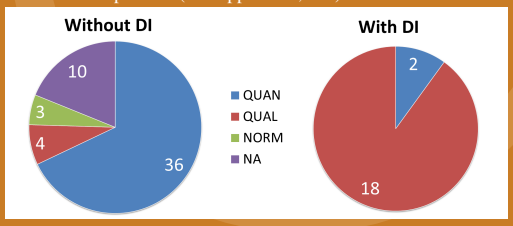
Patients with both discoloration and radiological signs of DI were diagnosed with DI.

Results

The distribution of OI types according to DI status of the patients is shown in the table below.

	OI I n=55	OI III n=6	OI IV n=12	Total 73
With DI	2 (4%)	6	12	20 (27%)
Without DI	53 (96%)	0	0	53 (73%)

The number of OI patients with or without DI according to the type of collagen defect: quantitatively deficient (QUAN), qualitatively deficient (QUAL), or normal (NORM). A skin biopsy was not obtained in 10 patients (Not applicable; NA).



Radiological signs of DI in the 20 OI patients with DI according to the type of collagen defect.

	QUAL n=18	QUAN n=2	TOTAL n=20
Obliterated pulp chambers	16	2	18 (90%)
Short roots	6	0	6 (30%)
Cervical constriction	13	0	13 (65%)

Adjunctive findings in the 20 OI patients with DI.
Dental agenesis (2 patients), impacted teeth (4 patients), extracted teeth (9 patients), endodontically affected teeth (5 patients), root fracture (2 patients), and root resorption (1 patient).

Conclusion

- The prevalence of DI (27%) was in the range of previously reported prevalence of DI in Scandinavian OI-patients (19%[1] - 42%[2])
- DI was a dominant finding in OI type III and IV
- DI was a rare finding in OI type I
- In patients with OI, DI was particularly associated with qualitative collagen defects
- Obliterated pulp chambers was the most pathognomonic finding of DI

References
1) Saeves R, Lande WL, Ambjornsen E, Axelsson S, Nordgarden H, Storhaug K. Oral findings in adults with osteogenesis imperfecta. Spec Care Dentist 2009; 29(2):102-108

2) Malmgren B, Norgren S. Dental aberrations in children and adolescents with osteogenesis imperfecta. Acta Odontol Scand 2002; 60(2):65-71

AARHUS UNIVERSITET

midt regionmidtjylland

Aarhus University Hospital DENMARK