Coll. Antropol. **30** (2006) 1: 231–234
Case report

Esthetic Reconstruction of Teeth in Patient with Dentinogenesis Imperfecta – A Case Report

Alena Knežević, Zrinka Tarle and Vlatko Pandurić

Department of Endodontics and Restorative Dentistry, School of Dental Medicine, University of Zagreb, Zagreb, Croatia

A B S T R A C T

Dentinogenesis imperfecta (DI) is the result of a dominant genetic defect and affects both the deciduous and permanent dentitions. It is characterized by opalescent teeth composed of irregularly formed and undemineralized dentin which obliterates pulp chamber and root canal. DI can appear as a separate disorder or with osteogenesis imperfecta (OI). The teeth with DI show a grayish-blue to brown hue with dislodged enamel, dysplastic dentine with irregular dentinal tubules and interglobular dentine, short roots and pulpal obliteration, which all may lead to rapid and extensive attrition which require adequate crown reconstruction. The aim of this study was to show a reconstruction of frontal teeth in upper jaw with direct composite veneers in young adult patient with DI.

Key words: *dentine, dentinogenesis imperfecta, osteogenesis imperfecta, teeth reconstruction*

Introduction

Dentinogenesis imperfecta (DI) is a hereditary defect consisting of opalescent teeth composed of irregularly formed and undemineralized dentin that obliterates the pulp chamber and root canals. DI may be present as a single disorder or in association with osteogenesis imperfecta (OI)[1]. OI ia an autosomal dominant disorder of connective tissue caused by mutations in the genes COL1A1 and/or COL1A2 that encode pro-α1 and pro-α2 chains of type I collagen which is the mayor protein of the organic matrix present in dentin and bone. OI is usually divided into four groups: type I, II, III and IV. Most cases of OI involve a dominant mutation. In OI, a dominant genetic defect causes one of two things to occur:

1. The dominant altered gene directs cells to make an altered collagen protein. Even thought the normal gene directs cells to make normal collagen, the presence of altered collagen causes type II, III or IV OI. These types result from a problem with the quality of collagen.

2. The dominant altered gene fails to direct cells to make any collagen protein. Although some collagen is produced by instructions from the normal gene, there is an overall decrease in the total amount of collagen produced, resulting in type I OI. This type results from a problem with the quantity of collagen. When a mutation is dominant, a person only has to receive one faulty gene to have a genetic disorder. This is the case with most people who have OI: they have one faulty gene for type 1 collagen, and one normal gene for type 1 collagen[1,2–4].

OI type III has been documented in some cases as recessive inheritance. Most researchers now agree that recessive inheritance rarely causes osteogenesis imperfecta[3–5].

Patients with OI may have blue sclera, hearing loss, growth deficiency, joint laxity, bone fragility, DI and also other dental abnormalities such as agenesis, apically extended pulp chambers, impaction, invagination, denticles[2,6–8].

In patients with DI teeth discolorations ranges from greyish-blue to brown and the dentin does not cushion the overlaying enamel adequately. The dentin is characterized by embedded cells, atubular, fibrous, irregular and interglobular dentine, while the thin peripheral layer (mantel dentin) is normally formed.

The enamel appears to be of normal chemically and histologically structure but tends to chip away from the dentin exposing the soft dysplastic dentine which can lead to rapid attrition as soon as they appear within the oral cavity. The dentino-enamel junction are smooth and not-scalloped.

The pulp chamber is usually obliterated and the pulp canals remains only as a thin slit or is also obliterated.

Received for publication March 10, 2005

The primary teeth are more severely affected than the permanent. Radiographically, the teeth show short roots, bulbous crown with constriction at the cervix of the crown and pulpal obliterations[9].

DI is divided into three groups: DI type I is afflicted with OI, type II is the most common and is known as a hereditary opalescent dentin, and type III (Brandwine type isolate opalescent dentin) is characterized by multiple pulpal exposure in deciduous dentition. The clinical, radiographic and histological manifestations of DI type I and DI type II are similar, although the clinical picture is more varied in DI type I. DI type II has been attributed to autosomal dominant mutations in the dentin sialophosphoprotein (DSPP) gene, encoding two dentin-specific non-collagenous matrix proteins, dentin sialoprotein (DSP) and phosphoprotein (DPP)[9,10]. DI type III is also attributed to autosomal dominant mutations and is characterized with specific shape of the teeth which is called »shell teeth«. The prevalence of all types DI is approximately 1:8000[10,11].

The rapid attrition of such teeth in patients with DI results very soon in a closed bite. The crowns should be reconstructed on deciduous and permanent molars as soon as they appear into the oral cavity, while even short delays result in wearing of the enamel crown to the gingival line.

The histological structure of the mantle dentin in DI as mentioned, appears relatively normal, but the scalloping at the dentinoenamel junction is decreased or missing. The scallopinghe mechanically lock of dentin and enamel together, and with its decrease or absence, the enamel fractures off easily. Therefore, the treatment of DI is focusing on protecting the affected dentin from caries, attrition, abrasion and erosion. The options for restorative treatment usually include crowns. Some authors recommend splinting the crowns while some other authors do not recommend using those teeth as abutments for crowns because of their brittenless[9,12,13].

Except in DI type II, endodontic procedure are usually unavailable for patients suffering from DI because of the obliteration of the root canals. If the canals can be found, endodontic treatment may proceed normally. However, in cases where endodontic treatment can not be done properly, post and core restorations my have questionable prognosis due to morphologic changes of the tooth structure and may lead to tooth fracture[14,15].

The use of overdentures on vital abutments for extremely worn dentitions or elderly patients may also be one of the restorative options.

The purpose of this clinical report is to describe a restorative treatment solution of young adult patient with DI.

Case Report

A 23 year old patient called the Department of Restorative Dentistry at the Zagreb School of Dental Medicine to have a dental examination (Figure 1). Prior to this the patient has been diagnosed OI with DI, but besides

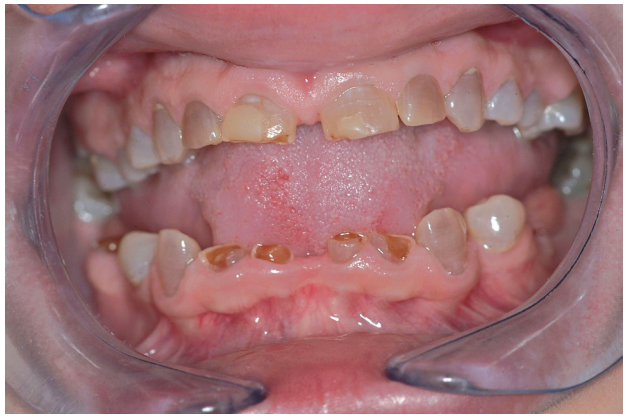

Fig. 1. Dental status of patient with dentinogenesis imperfecta.

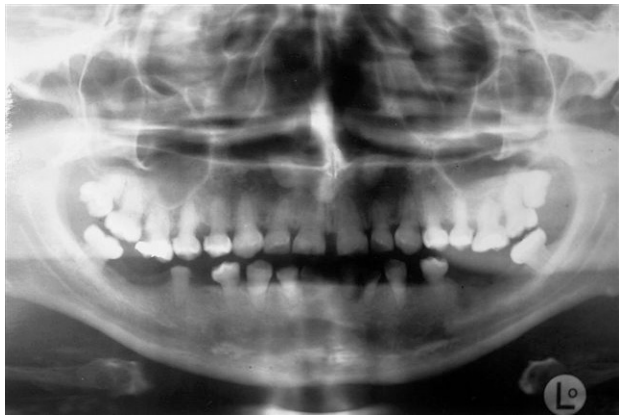

Fig. 2. Radiographic view (orthopantomogram) of patient with dentiongenesis imperfecta.

orthopantomogram (Figure 2) the patient did not have any other dental documentation. From his case history we find that the patient suffered from multiple fractures of lower limbs long bones before he reached 12 years of age which were treated with conservative and surgical methods. For a period of time he was confined to wheelchair but currently he can walk with the help of crutches. There was no family history of OI. By checking the oral cavity abrasion of upper and lower teeth was found. His lower teeth are almost completely abraded with the remains of dental cap at the gingival level. The condition of the teeth in the upper jaw was better; a lower level of abrasion was found occlusally, without significant caries lesion. Labial surfaces were slightly eroded, and the teeth were of yellow- brown color. Dental caps in the upper jaw were rounded, lowered in the cervical part. Between central incisor teeth there were diastemas (Figure 1). As we were dealing with a patient who was a refugee from an area affected by war, we decided to perform a correctional intervention in the upper jaw, by applying direct composite veneers from upper left canine to upper right canine in dental arch. As the enamel was well preserved, the upper enamel level was slightly angled with diamond bur. A classical procedure of applying composite veneers followed: transparent adapt strip was placed and

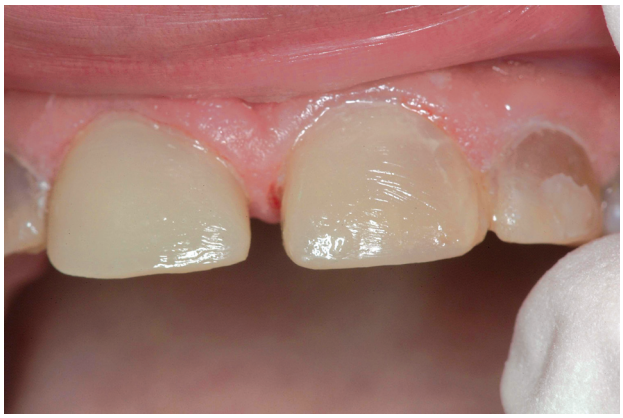

Fig. 3. Upper left and right central incisor after the reconstruction of labial surface with direct composite labial veneers.

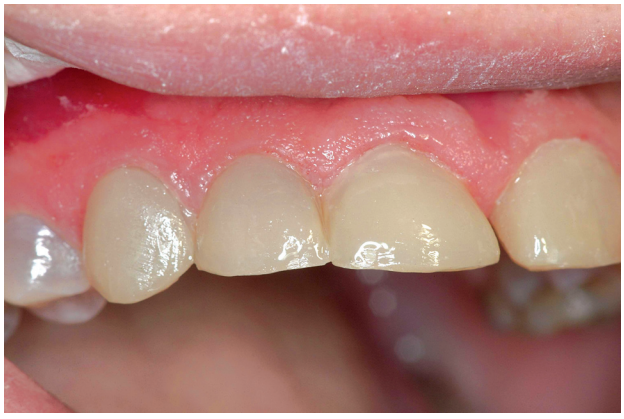

Fig. 4. Right upper frontal teeth (canine, lateral and central incisor) after the reconstruction of labial surface with direct composite labial veneers.

was fixed with interdental wedges mesial and distal side of the tooth. On the outer side the transparent strip was fixed with a bond to stop the secretion from gingival sulcus. The procedure of enamel etching by 37% ortophosphoric acid was carried out for 15 seconds. After that acid was rinsed, dried and adhesive was applied and polymerized. Then followed the placing of composite material or adequate shade in layers and curing of each layer separately. After curing, transparent strip and wedges were removed and the excess of composite resin were removed and veneers were modelled, finished and polished. Figures 3, 4 and 5 show final reconstructions of labial surface of upper teeth (from right upper canine to left upper canine). The diastema between upper central incisors was not completely closed because of the specific shape and size of central incisors.

Discussion

Esthetic is of a great concern especially in young patients and should be included in the plan of treatment.

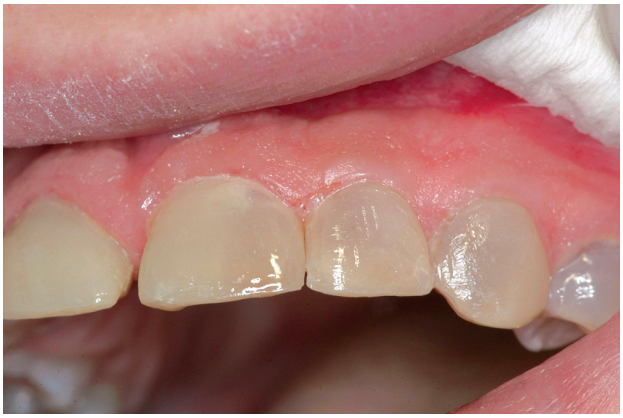

Fig. 5. Left upper frontal teeth (canine, lateral and central incisor) after the reconstruction of labial surface with direct composite labial veneers.

One of the properly solution is an composite veneers shown in this case report. The esthetic is satisfying and the teeth are minimally destroyed. For appropriate placement of composite resins and for the duration of composite restoration the preparation of hard tissue before composite placement is very important. The inorganic phase in DI dentin was investigated by Kerbel et al.[13] who reported that the crystallites in DI dentin, through of normal size are less numerous than in normal dentin. Electron microprobe analysis indicated significant differences in mineral content between DI dentin and normal dentin; in the former, Kerbel et all[13] observed a higher Ca/P ratio, an overall reduction in both Ca and P, and signifficantly less Mg. The decrease mineral content of DI dentin has been corroborated by chemical analyses[14].

Composite veneers are indicated in patients suffering from DI especially in frontal region in teeth which are without caries and with low abrasion. This treatment allowed optimal esthetic and function, as well as preserved the structure of the remaining natural teeth. In the cases where enamel is totally lost, self-etching dentine adhesives have advantage due to the lower mineral content in dentine. In contrary, total-etch of hard dental tissue before composite resin application, with shorter etching time (10–15 second) is indicated[15]. In this case total etch technique was used because enamel was mostly preserved[16,17]. Although, in theory, bonding to resin to the defective tooth structure may be compromised, it was clinically successful in most patients. For this reason, adhesive dentistry is not contraindicated N patients with DI. However, expectations of success with enamel/dentin bonding should be guarded, and the use of such treatment should be assessed on an individual bases because of the extreme variability of dentin involvement[1].

Conclusion

Composite veneers is one of the cheapest and fastest reconstructions that can be achieved with patients suffering from DI, especially with those who can not afford a

prosthetic substitute, as was the case with our patient. Composite veneers contributed to the improvement of the esthetic effect which in itself can also contribute to patient's psychosocial status.

Acknowledgements

This work was supported by Ministry of Science, Education and Sport, Grant No. 0065007, Zagreb, Croatia.

REFERENCES

1. O'CONNELL, Y. C., J. C. MARINI, Oral. Surg. Oral. Med. Oral. Pathol. Oral. Radiol. Endod., 87 (1999) 189. — 2. MALMGREN, B., S. LINSKOG, Acta. Odontol. Scand., 61 (2003) 72. — 3. CHEVREL, G., Osteogenesis Imperfecta, Orphanet encyclopedia, June 2004., accessed 28.10.2005. Available from: http://www.orpha.net/data/patho/GB/uk-OI-pdf. — 4. Osteogenesis Imperfecta Foundation: Genetics. OI Issues: Genetics, accessed 28.10.2005. Available from: http://www.oif.org/site/PageServer?pagename=Genetics — 5. OGUNSALU, C., B. HANCHARD, Austr. Dent. J., 42 (1997) 175. — 6. MALMGREN, B., S. NORGREN, Acta. Odontol. Scand., 60 (2002) 65. — 7. LUND, A. M., B. L. JENSEN, L. A. NIELSEN, F. J. SKOVBY, Craniofac. Genet. Dev. Biol., 18 (1998) 30. — 8. LINDAU, B., W. DIETZ, T. LUNDGREN, K. STORHAUG, J. G. NOREN, Int. J. Pediatr. Dent., 9 (1999) 253. — 9. MAYORDOMO, F. G., F. ESTERELA, E. A. DE ALDECOA, Quintessence. Int., 23 (1992) 795. — 10. XIAO, S., C. YU, X. CHOU, W. YUAN, Y. WANG, L. BU, Nat. Genet., 27 (2001) 201. — 11. ZHANG, X., J. ZHAO, C. LI, S. GAO, C. QIU, P. LIU, Nat. Genet., 27 (2001) 151. — 12. HENKE, D. A., T. A. FRIDRICH, S. A. AQUILINO, J. Prosthet. Dent., 81 (1999) 503. — 13. RIVERS, J. A., R. S. STAFFANOU, Compend. Contin. Educ. Dent., 6 (1985) 548. — 14. LEVIN, L. S., S. H. LEAF, R. J. JELMINI, J. J. ROSE, K. N. ROSENBAUM, Oral. Sur. Oral. Med. Oral. Pathol., 56 (1983) 267. — 15. PETTIETTE, M. T., J. T. WRIGHT, M. TROPE, C. HILL, Oral. Surg. Oral. Med. Oral. Pathol. Oral. Radiol. Endod., 86 (1998) 733. — 16. KERBEL, B., G. DACULSI, J. MENANTEAU, L. M. KEREBEL, J. Dent. Res., 60 (1981) 1655. — 17. WARING, D., M. S. DUGGAL, J. T. WRIGHT, J. Dent. Res., 74 (1995) 856. — 18. TAY, F. R., D. H. PASHLEY, N. M. KING, R. M. CARVALHO, J. TSAI, S. C. N. LAI, L. JR. MARQUENZINI, Oper. Dent., 29 (2004) 309. — 19. KEROS, J., I. CIGLAR, P. KOBLER, N. IVOŠEVIĆ, Coll. Antropol., 26 (2002) 651. — 20. AZINOVIĆ, Z., J. KEROS, D. BUKOVIĆ, A. AZINOVIĆ, Coll. Antropol, 27 (2003) 381.

A. Knežević

Department of Endodontics and Restorative Dentistry, School of Dental Medicine, University of Zagreb, Gundulićeva 5, 10000 Zagreb, Croatia
e-mail: ma505ak@yahoo.com

ESTETSKA REKONSTRUKCIJA ZUBI KOD PACIJENTA S DENTINOGENESIS IMPERFECTA – PRIKAZ SLUČAJA

SAŽETAK

Dentinogenesis imperfecta (DI) je rezultat dominantnog genetskog defekta i zahvaća mliječnu i trajnu denticiju. Karakterizirana je opalescentnim zubima građenim od iregularnog i nedostatno mineraliziranog dentina koji obliterira pulpne rogove i korijenski kanal. DI se može pojaviti kao zaseban poremećaj ili u sklopu osteogenesis imperfecta (OI). Zubi s DI pokazuju sivo plave do smeđe diskoloracije s nedovoljno mineraliziranom caklinom, displastičnim dentinom s iregularnim dentinskim tubulusima i inetrglobularnim dentinom, kratke korjenove s obliteracijom pulpnog prostora. Sve navedeno dovodi do brze i opsežne atricije koja zahtijeva odgovarajuću rekonstrukciju zubne krune. Svrha ovog rada bila je prikazati mogućnost rekonstrukcije frontalnih zubi gornje čeljusti direktnim kompozitnim fasetama u mladog odraslog pacijenta s DI.