Early Rehabilitation of Incisors with Dentinogenesis Imperfecta Type II – Case Report

Ana Paula CA Beltrame*/ Maíra Mery Rosa**/ Ricardo AT Noschang***/ Izabel CS Almeida****

Dentinogenesis imperfecta is an phenotypic alteration in the formation of the organic matrix. It causes the rapid and progressive wear of tooth structure, which may compromise tooth function and aesthetics. This is a case of a 1y, 8m-old child with dentinogenesis imperfecta. All teeth presented with an opalescent appearance and grayish color hue. Compromised structural integrity was noted as excessive wear and fracturing of the enamel from the dentin layer. With low doses of midazolam (oral) and chloral hydrate (rectal) administration, in a hospital environment, sedation was used to aid full mouth rehabilitation. Direct bonded restorations were performed on primary maxillary incisors and indirect restorations, pre-made on a plaster model using composite resin, were performed on primary mandibular incisors. After 32 months, we observed that diagnosis and early treatment allowed preventive maintenance of the patient's primary dentition, maintaining tooth function and esthetics.

Key words: dentinogenesis imperfecta, composite resin, resin veneers.

INTRODUCTION

Dentinogenesis imperfecta (D.I.) is a tooth formation disorder that affects 1/8000 individuals.[1] It is associated with an inherited genetic alteration of autosomal dominant origin with the mutation of chromosome 4q21.[2] This mutation alters the protein *dentin sialophosphoprotein* (DSPP) and its development, which generates a malformation in the organic matrix of the dentin.[2]

It affects both dentitions, being usually more intense in the primary dentition and can be classified into three types:[3] Type I, in addition to changes in the teeth, is associated with osteogenesis imperfecta; Type II is the most common, presenting the similar dental conditions but not associated with osteogenesis imperfecta; Type III is very rare and has been observed in a group with tri-racial miscegenation (Native American, African American and European Caucasian) in the Maryland region, in the US. In Type III, in addition to changes related to the color of the teeth, there are large pulp chambers with little calcification, pulp exposure, and generally shell teeth.

In dentinogenesis imperfecta, tooth enamel usually develops properly, however, the enamel-dentine junction is smooth, wrinkle-free, unlike what is considered the norm.[4,5] This characteristic favors enamel detachment and fractures, exposing the underlying malformed dentin.[5]

Below the amelo-dentin junction, due to structural malformation, the dentin appears altered, with low mineralization, irregularities, few and often obliterated dentinal tubules, causing higher fragility.[6] The exposed dentin wears quickly, leading to a reduction in the vertical dimension, pulp exposure and even root fracture under load, since the roots are narrow and slightly mineralized.[4,5,7] This change in dentin mineralization also affects the color of the tooth.[6]

Individuals with this phenotype clinically present teeth color change ranging from blue-gray to yellowish brown and increased opalescence.[1,4] Radiography shows bulbar crowns, constriction in the cervical region of the teeth and narrow roots.[2,5] In initial assessments, individuals may present increased pulp chamber, which later suffers a process of obliteration due to dentin deposition.[7]

Through discoloration and abnormal wear patterns, this phenotypic alteration is a chronic, progressive degradation of the child's dentition that, typically, negatively influences the child's peer/social relationships.[4]

In addition to the functional and aesthetic results, the treatment prevents significant wear of the tooth and the possibility of tooth loss.[5] Thus, the diagnosis should be made as early as possible, as well as the implementation of treatment.[4]

From the Department of Dentistry, Graduate Program in Dentistry, Concentration Area–Pediatric Dentistry, Federal University of Santa Catarina, Florianópolis, Santa Catarina, Brazil.
*Ana Paula C A Beltrame, DDS, MS, PhD student.
**Maíra Mery Rosa, DDS.
***Ricardo A T Noschang, DDS, MS, PhD student.
****Izabel C S Almeida, DDS, MS, PhD, Full Professor.

Send all correspondence to:
Ricardo Augusto Tomaz Noschang
Elizeu di Bernardi 200, loja. 06
Zip Code: 88101-050
Phone: 55 48 9106 9042
E-mail: ricardo.noschang@posgrad.ufsc.br

In the case of children, in addition to the actual clinical procedures, there is a need to manage behavioral cooperation because this is generally a complicating, limiting factor.

Several treatments for improving oral conditions have been proposed, including direct bonded composite restorations, veneers and indirect fabricated composite resin restorations, as well as, dental implants and full mouth rehabilitation with partial or full dentures.[2,4]

The objective of this paper is to present the diagnosis of a 1 year, 8 month old pediatric patient of D.I. and the subsequent treatment using direct and indirect composite resin restorations and the associated follow-up care.

Case Report

The patient LML, 1-year, 8-month-old, attended the Pediatric Dental Clinic of the Federal University of Santa Catarina, in Florianópolis, Brazil. The patient's mother, accompanied her daughter at the initial exam and reported that her daughter's teeth began to break as soon as they erupted, subsequently presenting with sensitivity while ingesting cold foods. In her history, it was found that the child had good health with no significant history of disease. Upon clinical examination, an incomplete primary dentition with absence of the primary second molars was observed. The remaining teeth had an opalescent sheen with a grayish hue. All remaining teeth showed incisal wear and fractures into the dentinal layer; however, the primary mandibular incisors had significantly more coronal tooth wear (Figure 1). Clinically, these signs were consistent with the diagnosis of dentinogenesis imperfecta. No carious lesions were observed and parent reaffirmed the child's healthy food and hygiene habits. Upon conducting an oral clinical examination of the patient's mother, highly similar opalescent-grayish sheen was observed on her remaining natural teeth with the majority of her other teeth having partial-full coverage restorations. The treatment plan was organized in three subsequent phases (prophylaxis, restorative, and follow-up care). In order to further reduce the sensitivity until the restorative procedures were defined and implemented, the first treatment phase included prophylaxis with an application of fluoride varnish. A total of four visits to apply fluoride varnish and proper prophylaxis was required. The second treatment phase required four separate appointments to clinically apply the planned restorations of choice. The anterior primary maxillary incisors lacked significant wear and direct restorations with composite resin were planned. Whereas, due to the significant loss of viable dental structure and possibly enabling greater longevity of the treatment for the primary lower incisors, the treatment option selected was for indirect composite resin restorations (prefabricated on a plaster stone model). After completion of the restoring procedures, two quarterly follow-up visits were also scheduled.

During treatment, due to the young age of the patient and the need to control her behavior, the dental team opted for a medical anesthesiologist to administer sedation for all restorative treatment appointments. Informed consent was obtained from parents of the child for the treatment described.

The first phase of the clinical treatment was performed in a dental office, with immobilization of the child by her mother, so that the prophylaxis consultations and topical application of 5% fluoride varnish (Duraphat®, Colgate-Palmolive Ltda, São Paulo, Brazil) could be performed (Figure 2). Subsequently, the patient was admitted in a hospital environment at the University Hospital of the Federal University of Santa Catarina, under sedation with an initial oral dose of midazolam (0.3 ml/Kg) and followed by a rectally administered chloral hydrate (1 ml/Kg, enema 10%), performed and managed by the medical anesthesiologist, and the direct restorations of the primary maxillary incisors (51,52,61,62) with composite resin, were carried out by two pediatric dental residents. Since there was no need for tooth preparation or additional retention, local anesthesia was not required or used. The teeth received prophylaxis, etching and application of adhesive (Single Bond, 3M of Brasil, Brazil). The restorations were made using acetate crowns (TDV, Pomerode, Brazil) and relative isolation. Acetate crowns were tested, adjusted, drilled for draining excesses and filled with composite resin (Charisma, Heraeus Kulzer, Hanau, Germany) in color A1 (Figure 3). They were adapted to the teeth and light-cured on the labial and lingual surfaces for 40 seconds each. For the reconstruction of teeth 71, 72, 81 and 82, an addition silicone (Express XT, 3M ESPE, Germany) model was made in a dental office, with a double dose of the catalyst, along with a special stone plaster working model. With the aid of the plaster-working model, restorations were pre-made with composite resin (Charisma, Heraeus Kulzer, Hanau, Germany), also in color A1 (Figure 4), incrementally fabricated to ensure proper UV light polymerization. In the following appointment, with the patient under sedation as previously described, the lower incisors were prepared with prophylaxis, etching, application of adhesive agent (Single Bond, 3M do Brasil, São Paulo, Brazil), and ultimately cemented to lower anterior incisors with resin cement (RelyX, 3M ESPE, Hanau, Germany). The final restorative appointment was held in the dental office, where the indirect restoration's occlusion was adjusted and provided a final polish (Figure 5).

The patient was seen after three months for evaluation of the restorations and observation of the erupting natural teeth's condition. In the second evaluation, after six months, the restorations were intact and the deciduous dentition had completely erupted, with no clinically observable loss of molars and/or canines. After 32 months of observation the restorations continue to play it's role with success (Figure 6).

DISCUSSION

In this case of dentinogenesis imperfecta where an pediatric patient age <2 years old, the current early diagnosis and treatment was to manage the process of tooth wear that will likely progress to teeth sensitivity, pulp damage/exposure, and problems related to the change of the vertical dimension. Additionally, as the degradation, wear, and discoloration of teeth rapidly progress cases there is an even greater opportunity/need to treat and restore aesthetics and function.[5] This aesthetic concern observed in most cases of D.I. have a high correlation to negatively patient's self-esteem, additionally damaging their interpersonal relationships, and is directly linked to their quality of life, regardless of the age range in which the patient is included.[4]

Previously, the indicated treatment of dentinogenesis imperfecta was to postpone the intervention until the beginning of the permanent dentition.[4] This orientation, however, caused the need for major rehabilitation. Delgado et al. cited that there is no formal protocol as for the ideal age for treatment, suggesting that a thorough analysis of each case should be made, taking into account the stage of teeth wear.[5] However, Bouvier et al, emphasized the need for treatment

Figure 1. Initial aspect of the patient, with wear in the anterior incisors.

Figure 2. Application of fluoride varnish.

Figure 3. Reconstruction of tooth 51.

Figure 4. Restorations completed on the working model.

Figure 5. Appearance of completed composite resin restorations.

Figure 6. 32 months follow up

when the first signs of loss of structure resulting from this disorder were observed.[4] In most cases, understanding the typical rampant dental wear and subsequent loss of the dentition associated with D.I., which will likely require complex restorative needs like dental implants and full arch prosthetics, further supports the need for earlier diagnosis and interceptive treatment. Sapir et al. cite as other advantages of early intervention the maintenance of arch size and normal growth of the facial bones.[7]

In the primary dentition, the most common treatment is the reconstruction of each element with dental restorative materials and crowns or the making of partial or full dentures, in some situations using the affected teeth as support.[4,6] In this clinical case, as the teeth offered sufficient support, the primary maxillary incisors were restored directly and the primary mandibular incisors were reconstructed indirectly (both using composite resin as the restorative material of choice). Sapir et al. also opted for composite resin to rehabilitate anterior teeth in patients up to 24 months old.[7]

Despite the frequent use of this material, the quality of adhesion to enamel and dentin in cases of D.I is questionable.[8] There is the assumption that there is no adequate formation of the hybrid layer and this would compromise the quality and life span of restorations. According to Leal et al., the decision for the composite resin technique should be restricted to mild to moderate cases.[6]

In order for restorative procedures to be properly performed, it was necessary to control the patient's behavior using sedative agents. A licensed medical anesthesiologist administered an initial oral dose of midazolam, followed by a rectally administered dose of chloral hydrate, substances that synergistically promote a longer working time and more predictable sedative control of the patient. Both were proven to be clinically safe for use on children and efficacious with few side effects in case of short-term sedation.[9] In the upper teeth, the use of acetate crowns made the implementation of restorations quicker and easier, considerably reducing clinical time, favoring the sculpture of teeth and a good cosmetic result.[10] In the mandibular arch, due to the significant loss of structure, small size in the mesial-distal direction and shape of the crowns, the restorations were performed with composite resin, but through the indirect technique.

The patient's mother was examined and also had dental characteristics of dentinogenesis imperfecta. Since D.I. is a genetic mutation, family members such as grandparents, uncles and cousins can present phenotypic expression of this disease as well.[2] Patients diagnosed with dentinogenesis imperfecta and their families should be informed of the possibility of transferring this genetic abnormality to future generations.[5]

After the fifth follow-up visit (thirty two months later), all restorations remained intact, promoting function and aesthetic maintenance, as well as, preventing further wear of tooth structure, pulp damage and the other various complications. We emphasize the importance of early treatment, which promotes a favorable prognosis and the need for regular monitoring of the patient for the maintenance of the results.

CONCLUSION

This case report is an example that correct diagnosis and early treatment can provide dental function and esthetics in a very young patient affected with dentinogenesis imperfecta, maintaining and protecting the patient's primary dentition with long term results.

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