

Dentinogenesis imperfecta type II: a case report of a 34-year follow-up

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ABSTRACT | Dentinogenesis imperfecta (DI) is a hereditary developmental disorder of dentin formation that can occur associated with osteogenesis imperfecta (type I), isolated (type II), or in a specific isolated resident group of Brandywine, in southern Maryland (type III). This work aims at reporting a clinical case of DI type II in childhood with a 34-year follow up. The child at issue was taken to the dental health service at a very young age, which favored an appropriate treatment, avoiding complications, and portending a favorable long-term prognosis, besides safeguarding the patient physical and mental well-being. The clinical aspects of this condition are teeth with short crowns and gray-brown coloration, and an altered consistency of affected dental elements. Radiographically, the teeth present bulbous crowns, cervical constriction, thin roots, and early obliteration of the root canal and pulp chambers due to excessive dentin production. Rehabilitation treatment included the use of stainless-steel crowns for reconstructing deciduous molars and composite resin restorations on the anterior deciduous teeth. As for permanent dentition, it consisted of aesthetic-functional rehabilitation using metal crowns on the first molars and ceramic crowns and facets on the anterior teeth. Endodontic, prosthetic and restorative treatment was performed on other posterior teeth. Preventive measures were instituted. DI may cause serious changes in dentin structure, affecting function and aesthetics in both dentitions. The sooner it is administered, the more promising the multidisciplinary dental treatment will be in promoting health and minimizing damage to affected individuals.

DESCRIPTORS | Dentinogenesis Imperfecta; Early Diagnosis; Mouth Rehabilitation; Continuity of Patient Care.

RESUMO | **Dentinogênese imperfeita tipo II: um relato de caso com acompanhamento de 34 anos** • A dentinogênese imperfeita (DI) é uma doença de alteração hereditária que afeta o desenvolvimento dentinário, podendo ocorrer associada à presença de osteogênese imperfeita (tipo I), isoladamente (tipo II), ou especificamente associada ao povoado de Brandywine, no sul de Maryland (tipo III). O objetivo deste trabalho é relatar um caso clínico de DI tipo II na infância que recebeu acompanhamento por 34 anos. A criança em questão foi encaminhada ao serviço de assistência odontológica ainda muito pequena, o que favoreceu um tratamento adequado, evitando complicações e trazendo um prognóstico favorável a longo prazo, além de garantir o bem-estar físico e mental da paciente. Os aspectos clínicos desta condição são dentes com coroas curtas e coloração marrom acinzentada, além de uma consistência alterada nos elementos dentários afetados. Radiograficamente, foram identificados dentes com coroas bulbosas, constrição cervical, raízes afinadas, e obliteração precoce do canal radicular e câmaras pulparez devido à excessiva produção de dentina. Para o tratamento, utilizou-se coroas de aço cromado na reconstrução de molares decíduos e restaurações de resina composta nos dentes decíduos anteriores. Quanto à dentição permanente, o tratamento visou reestabelecer a função e a estética, utilizando-se de coroas metálicas nos primeiros molares e coroas e facetas cerâmicas nos dentes anteriores. Em outros dentes posteriores foram utilizados os tratamentos endodôntico, protético e restaurador. Medidas preventivas foram instituídas. Em conclusão, a DI pode causar sérias alterações na estrutura dentinária, afetando a função e a estética de ambas as dentições. A intervenção odontológica multidisciplinar será mais promissora quanto mais cedo for estabelecida, promovendo a saúde bucal e minimizando os danos aos indivíduos afetados.

DESCRITORES | Dentinogênese Imperfeita; Diagnóstico Precoce; Reabilitação bucal; Continuidade do Atendimento ao Paciente.

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INTRODUCTION

Dentinogenesis imperfecta (DI) is a tooth developmental disorder associated with an alteration in organic matrix formation that may affect both dentitions, but occurs mostly in deciduous teeth. It can be classified into three types:¹ in type I, teeth changes are associated with osteogenesis imperfecta; type II entails dental features similar to those of type I, although unrelated to osteogenesis imperfecta and more common;² type III is very rare and observed within a tri-racial isolated group (Native American, African American, and European Caucasian) in the Maryland region.³

In DI type II, changes are related to mutations in dentin sialophosphoprotein (*DSPP*) gene. Teeth appear amber and opalescent, varying from brownish to bluish-gray, and pulp chamber is obliterated by abnormal dentin.^{2,4} Although unaffected, the enamel tends to fracture and easily detach due to abnormal amelodentinal junction, which causes dentin to undergo severe and rapid attrition and leads to a marked shortening of the teeth. Radiographically, teeth presents bulbous crowns, resulting from cervical constriction, with thin and short roots. Initially, pulp chambers may be abnormally wide and resemble “shell teeth,” but they will progressively obliterate.¹

The treatment of this condition is difficult and complex, and should entail multiple interventions – from hygiene and nutritional orientations to dental and skeletal rehabilitation, when necessary¹ – and be initiated as soon as possible to minimize damage.^{3,4} Applying topical fluoride may be of great help in decreasing sensitivity and preventing caries.¹ In more severe cases evolving significant enamel fractures and rapid tooth wear, the treatment of choice is the use of complete aesthetic or metal crowns.^{1,5} Overdenture therapy should be considered in cases with severe coronary structure loss.⁶

This study aimed to report a clinical case of a DI type II patient with a 34-year follow-up, addressing clinical, radiographic, and restorative/prosthetic aspects to stress the importance in diagnosing and administering a rehabilitation treatment for patients physical and mental well-being.

CASE REPORT

A 2-year-old female patient (F.S.S.P.), from Ribeirão Preto (SP, Brazil), was referred for dental evaluation at the Pediatric Dentistry Clinic of the School of Dentistry of Ribeirão Preto, at the University of São Paulo. She mainly complained about her teeth being worn and dark (Figure 1a).

According to her guardian, the patient had already undergone several treatments without success. During anamnesis, we verified the absence of any systemic disease and family history of the condition. The child presented physical characteristics of normality, and no disorder and bone fracture, so we ruled out osteogenesis imperfecta. The mother also reported that the child had never used tetracycline.

Clinical examination revealed that the patient was at deciduous dentition phase and all teeth presented grayish-brown color, compromising esthetics. We also verified wear in the occlusal surfaces of deciduous molars and incisal surfaces of the upper deciduous incisors and canines, causing vertical dimension loss (Figure 1b).

For oral rehabilitation, deciduous molars underwent dental restoration with stainless-steel crowns to restore vertical dimension lost by excessive wear (Figure 1c). Anterior deciduous teeth underwent composite resin restorations to reestablish aesthetics. Treatment also entailed prophylaxis, topical fluoride applications, supervised tooth brushing, and nutritional orientations. During mixed dentition phase, the first permanent molars received resinous sealant and the patient was followed.



Figure 1 | Clinical appearance at deciduous dentition phase. (a) Superior occlusal view, showing teeth wear and color alteration. (b) Front view, showing aesthetic compromise and dental surfaces wear. (c) Frontal and inferior occlusal views, showing chrome steel crowns placement in deciduous molars.

During permanent dentition phase, clinical examination verified that all teeth presented a grayish-brown coloration, compromising aesthetics. It also detected occlusal surface wear on the upper and lower first molars and the upper premolars, causing

vertical dimension loss. The incisal edge of the anterior permanent maxillary and mandibular incisors and canines also presented wear (Figure 2a). However, the patient reported feeling no pain or dentin sensitivity and showed good periodontal health.

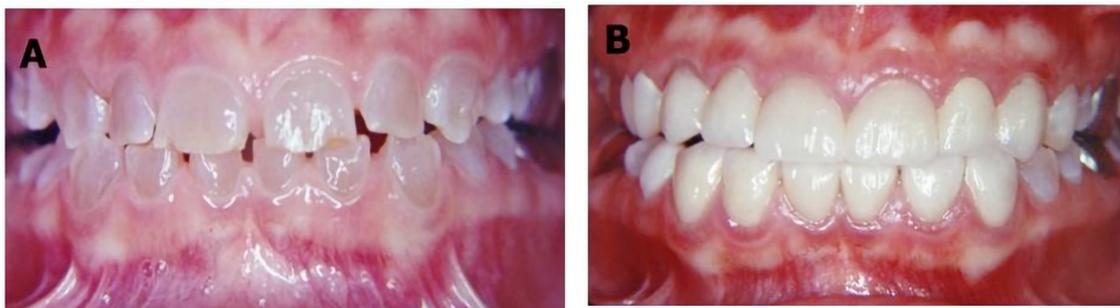


Figure 2 | Clinical aspect at permanent dentition phase. (a) Front view, showing areas of grayish-brown coloration and dental wear. (b) Front view of completed rehabilitation treatment, showing the excellent aesthetic result obtained.

Radiographic examination revealed obliteration of pulp chambers and root canals in all teeth, common in cases of DI type II (Figure 3). The new treatment plan included all-ceramic crowns on the lower anterior teeth, full metal crowns on the upper and lower first molars, and ceramic laminate veneers on the upper anterior teeth. We instituted

preventing measures considering the patient's caries risk, including oral hygiene, nutritional orientation, and topical fluoride application. The upper first premolars, left upper second premolar, and right lower second molar also underwent restorative treatment. The right upper second premolar received endodontic treatment, cast metal core, and ceramic

crown, and the left lower second molar received endodontic and restorative treatment. Figure 2b shows the finished prosthetic treatment.

After treatment completion, the patient attended no follow-up appointments. Recently, we contacted the patient to schedule another

appointment. Figure 4a shows the clinical appearance of the patient's permanent dentition at 10 years after treatment initiation. Figure 4b shows the current oral condition at 34 years after treatment initiation, evidencing the success of the measures instituted.



Figure 3 | Periapical radiographs at permanent dentition stage, showing the thin and short aspect of roots and obliteration of pulp chambers and root canals in all teeth.



Figure 4 | (a) Clinical appearance 10 years after treatment initiation; (b) Clinical appearance 34 years after treatment initiation.

DISCUSSION

Treating dentinogenesis imperfecta (DI) can be challenging, but the soon it starts, the greater are the chances of achieving success and reducing dental losses. According to Gama et al.,⁷ the disease hereditary nature may help in the early diagnosis and the development of a treatment plan. In our case report, the child had no family record with similar dental abnormalities. Yet, she was taken at a very young age to the dental health service, favoring an adequate treatment, protecting deciduous dentition from possible complications, such as wear and pulp injuries, and portending a favorable long-term prognosis.³

For treating mixed and permanent dentition, it is important to adopt a multidisciplinary approach that entails collaboration among dental clinicians, pediatric dentists, and orthodontists, focusing on prosthodontists.⁸ The literature addresses different treatment approaches, including outpatient procedures under general anesthesia, especially in severe cases. Parents and children motivation in following oral hygiene and nutritional instructions is also important to achieve the expected results in all cases of DI treatment.⁹

Performing stainless-steel crowns in deciduous or mixed dentition is recommended as a temporary treatment to protect the teeth while waiting for complete dentition replacement, bone growth, and vertical dimension reestablishment.^{4,10} Our patient underwent stainless-steel crowns on the deciduous posterior teeth and showed satisfactory results, fulfilling all purposes.

Full-coverage restoration is indicated in cases diagnosed with extensive tooth wear.⁷ The literature also recommends stabilizing the occlusion before an excessive loss of vertical dimension. However, in cases with a significant vertical dimension loss, the preconized treatment should reestablish it.¹ Considering that, the chosen treatment to reestablish vertical dimension for permanent dentition during dental rehabilitation consisted of metallic and

ceramic crowns, large composite resin restorations, and ceramic veneers.

After 24 years, we observed that ceramic crowns, metallic crowns, and ceramic laminates veneers were maintained and vertical dimension preserved. This indicates that, although lower for aesthetic reasons, the use of metal compounds in treatment has higher durability and wear resistance. It also avoids unscheduled returns and consultations, reducing biological contamination risks for both patient and dental team, especially in pandemic situations.

The treatment of dentinogenesis imperfecta varies according to each case specific severity.⁹ Regardless of the current aesthetic concern evolving even the posterior teeth,¹¹ the treatment plan here presented is a functional, effective, and economically viable option for DI patients.

Despite the need for new aesthetic rehabilitation plan, our patient was satisfied with the performed treatment. The present clinical case was evidently successful: even with unfavorable root anatomy, the treatment provided a high durability of the established prosthetic rehabilitation and preserved dental structures, proving to be adequate.

Understanding DI, its characteristics, and possible sequelae is of vital importance to enable a recognition of its clinical signs and an early implementation of preventive and restorative/prosthetic procedures for improving patients' quality of life.

CONCLUSION

The main goals of the treatment of DI patients are to prevent dental wear, restore aesthetics and, when necessary, restore occlusion. An early diagnosis ensures an early implementation of preventive and restorative and/or rehabilitation measures, providing aesthetic and functional satisfaction, minimizing dental loss, and improving the patient's quality of life. Quality treatment with an interdisciplinary approach and continuous follow-up can lead to long-term outcomes for the patient.

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