

CASE REPORT

Management of Dentinogenesis Imperfecta: A Report of Two Cases

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ABSTRACT

Dentinogenesis imperfecta is a hereditary dentin defect leading to discoloration as well as early tooth wear. Timely diagnosis and treatment are required to prevent further tooth loss. Two patients reported to the department of pediatrics dentistry with dentinogenesis imperfecta (DI). In the first case, patient complained of discoloration of both primary and permanent teeth and delayed eruption of permanent teeth. In the second case report, patient reported with mobile anterior tooth and missing teeth due to extraction of mobile teeth. Extraction was followed by space maintainers in the first case, and extraction was followed by partial denture in the second case. Both the patients and their parents were happy with the treatment provided.

Keywords: Dentin defect, Dentinogenesis imperfecta, Shell teeth.

International Journal of Clinical Pediatric Dentistry (2019): 10.5005/jp-journals-10005-1681

INTRODUCTION

Dentinogenesis imperfecta is a hereditary dentin defect caused by an autosomal dominant mutation in dentin sialophosphoprotein (DSPP) gene.¹ Dentinogenesis imperfecta is a localized mesodermal dysplasia affecting primary as well as permanent dentitions.² Dentinogenesis imperfecta is the most common genetic dental defect, approximately affecting 1 in 8,000 births in the USA.³ The cause of DI is mutation of chromosome 4q21 that alters the DSPP and its development, generating a malformation in the organic matrix of dentine and primary defects in the calcified matrix.⁴ The earlier classification given by Shield categorized DI into 3 types—type I: DI associated with osteogenesis imperfecta; type II: teeth having the same clinical, and radiological, and histological findings as DI type I, but is not seen with osteogenesis imperfecta; and type III: DI of a rare type seen in the Brandywine triracial population from Maryland and Washington, DC, USA. As evidence shows that there are 2 different entities, classification is devised into DI-I and DI-II, where DI-I is same as that of Shield's type II and DI-II is same as Shield's type III.⁵

The diagnosis of DI involves a thorough family history and detailed clinical and radiographic examination. In DI-I, all teeth in both dentitions are affected. Primary teeth are affected more severely, followed by the permanent incisors and first molars, with the second and third molars being the least-altered. The tooth color change is ranging from blue-gray to yellowish brown and increased opalescence. An amber translucent tooth color is common. Although the enamel structure is histologically normal, it tends to break. The dentin enamel junction is not interwoven to the normal extent; therefore, it breaks easily. The exposed dentine may subject to severe and rapid attrition. Radiographically, pulp chamber obliteration can be seen even before tooth eruption demonstrating "shell teeth" and multiple pulp exposures, whereas, in DI-II, the predominant characteristic is bell-shaped crowns, especially in the permanent dentition. Unlike type I, "shell teeth" demonstrate normal-thickness enamel in association with extremely thin dentin and dramatically enlarged pulps. The thin dentin may involve the entire tooth or be isolated to the root.⁶

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How to cite this article: Kaur A, Kumar S, Karda B, *et al.* Management of Dentinogenesis Imperfecta: A Report of Two Cases. *Int J Clin Pediatr Dent* 2019;12(5):464–466.

Source of support: Nil

Conflict of interest: None

Dentinogenesis imperfecta may require a multidisciplinary approach involving restorative, prosthodontic, and orthodontic treatments. The options for pediatric dentist for dental treatment include a stainless steel crowns (SSC), a simple removable space maintainer appliance, and adhesive restoration. These case reports present the clinical and radiographic findings along with the treatment given as space maintainer with SSC and prosthetic rehabilitation.

CASE DESCRIPTION

Case 1

A 10-year-old patient reported to the Department of Pediatric and Preventive Dentistry with the chief complaint of discoloration of teeth and worn-out upper and lower teeth. Clinically, 16, 55, 54, 53, 11, 21, 63, 64, 65, 26, 31, 32, 74, 35, 36, 41, 42, 83, 84, 85, 46 were present. Parents were concerned about worn-out teeth and yellowish discoloration.

The patient's family history revealed that her 5-year-old brother and mother have similar dental problems in their dentition. Parents had no consanguineous relationship. The color of her permanent teeth was bluish gray, whereas primary teeth were yellowish brown in color with attrition (Figs 1 and 2). Radiographic examination revealed bulbous crowns, obliteration of pulp chambers, lack of pulp horns, and constricted short roots.



Fig. 1: Preoperative maxillary area



Fig. 2: Preoperative mandibular area



Fig. 3: Lingual arch cemented after extraction of teeth

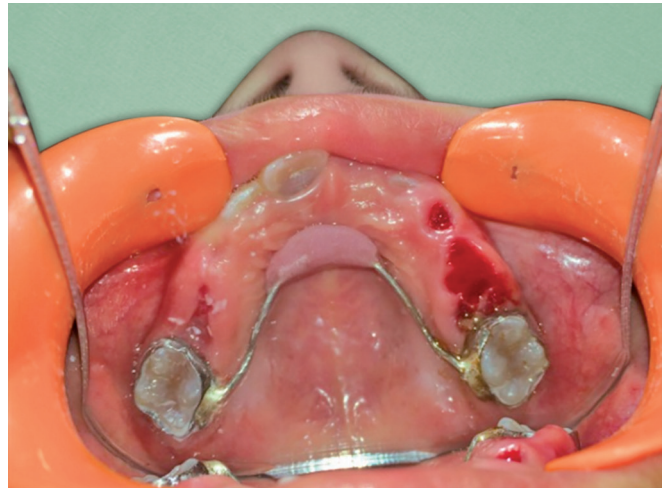


Fig. 4: Nance palatal arch space maintainer cemented after extraction of teeth



Fig. 5: Second case showing missing permanent teeth

Objective of treatment was protection to the posterior teeth from tooth wear and extraction of worn out primary teeth to accelerate the eruption of succedaneous teeth in buccal segment followed by space maintainer. In the first visit, oral prophylaxis, composite restoration of 35, and alginate impressions of both the

arches were taken. In the next visit after 2 days, band adaptation was done on 16 and 26, and extraction was done in maxillary and mandibular arch (53, 54, 55, 83, and 84) patient was given postoperative instruction and made to wait for half an hour till bleeding stopped. In third appointment, band adaptation on 36 and 46 and later extraction of 64, 65, and 75 were done. After half an hour, patient was checked and lingual arch (Fig. 3) and Nance palatal arch (Fig. 4) space maintainers were delivered.

Case 2

A 14-year-old patient reported to the department with a chief complaint of loose upper front tooth and 2 missing front teeth and 2 missing first permanent molars (Fig. 5). The main concern of the patient's parents was esthetics as two of the upper front teeth had already been extracted, owing to their high mobility. The oral examination showed a total loss of 4 teeth. The color of the remaining teeth showed an amber translucent hue. Further examination showed mobility with respect to 21 and a root stump with respect to 22. The 26 showed caries requiring a composite build up. Also seen was the peculiar mesial rotation of the mandibular second premolars. The orthopantomograph showed teeth with short roots and large pulp chambers. His younger sibling too had DI. Alginate impressions of the upper and lower arches were made



Fig. 6: Maxillary and mandibular removable partial denture

and casts poured. Then, the composite restoration of 26 was done. Two teeth, 21 and 22, were extracted, and patient was sent home with post-op instructions. Heat cured immediate upper (11, 21, and 22 regions) and lower removable partial dentures (RPDs) (36 and 46 regions) were fabricated and delivered to the patient on the recall appointment (Fig. 6). After this, it was made sure that the patient had no problems with his upper or lower RPDs, thereby restoring mainly the esthetics as well as the lost function. The patient is being followed up every 3 months to check the prosthesis as well as his oral status.

DISCUSSION

Fundamental defect in DI is mesodermal in which primary structural abnormality is in dentin. Kerebel et al.⁷ and Wright et al.⁸ showed gross abnormality of dentinal tubules and dentinal calcification, whereas enamel, cementum, and periodontal ligament are normal. The affected dentin has less of calcium (Ca), phosphorous (P), magnesium, a higher Ca:P ratio, and higher water content. The chief characteristic of DI-I is higher where rates due to the absence of intrafibrillar mineralization.⁹ Genetically, DI-1 is transmitted as an autosomal dominant Mendelian trait with almost 100% penetrance. Family history was positive in both the cases, and parents were informed about the possibility of transferring to the next generation in future.

Unwelcomed tooth wear, sensitivity, delayed eruption of teeth, discoloration and extensive tooth decay are common in DI. In mixed dentition, additional care is provided to preserve the arch perimeter. Early diagnosis and treatment are necessary to protect esthetic and function in patients with DI. Dental caries are prevented and treated at the initial stages so as to prevent further deterioration of the dentition. The patient's age, degree of carious involvement, patient cooperation, and dentition period are factors taken into consideration. Stainless steel crowns are recommended for both the primary and permanent dentition to minimize problems due to excessive tooth wear.

For discoloration, a variety of treatments are available, e.g., vital bleach, veneers, dental implants, overdenture prostheses, SSC, metal ceramic restorations, and adhesive restorations.

In the first case, the intention was to manage delayed eruption of permanent teeth and extensive primary tooth wear. As no set guidelines are available on the management of DI, customized treatment plan is recommended to reduce the further loss of tooth material and to take care of developing permanent dentition. To speed up the eruption of succedaneous teeth, primary teeth were extracted. Nance palatal and lingual arch space maintainers were planned to hold the space until the eruption of permanent teeth. Initially, bands were placed on first permanent molars for space maintenance. As patient was not able to maintain oral hygiene, SSC were given to prevent decalcification of these teeth.

In the 2nd case, we provided upper and lower RPDs to compensate multiple tooth loss, with the aim of restoring the patient's main concern—esthetics. In DI, the deterioration of the enamel–dentin junction results in the ready loss of enamel. The main goals of DI treatment are to protect the dentin from caries and to prevent tooth attrition, abrasion, and erosion. The dental treatment of patients with DI should protect the structure, esthetics, and tooth function. Also, prosthetic crown restoration could be considered in esthetic treatment planning in young patients.¹⁰

CONCLUSION

The main issue with DI patients is the prevention of further tooth material loss and restoration of oral function. In young children, esthetic treatment becomes more important to avoid peer pressure. Both the patients were satisfied with the treatment provided.

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