Case Report

Indian Journal of Forensic Odontology
Volume 17 Number 2, July - December 2024
DOI: http://dx.doi.org/10.21088/ijfo.0974.505X.17124.5

Management of Dentinogenesis Imperfecta by Prosthodontic Approach: A Case Report

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How to cite this article:
Anas Khan, Gibi Babu Philip, Ann Mary Varghese *et al.* Management of Dentinogenesis Imperfecta by Prosthodontic Approach A Case Report. Ind J Forensic Odontol 2024;17(1):79-82.

Abstract

Dentinogenesis imperfecta (DI) is a genetic disorder affecting the developmental integrity of the dentin which results in weakened dentin. It is inherited as an autosomal dominant trait. The affected teeth, generally the posterior teeth will have to be extracted due to severity in wear and thereby possible fracture. This commonlyresults in loss of posterior occlusion and reduced vertical dimension. Apart from wear and fracture, anterior teeth may have an unesthetic appearance because of discoloration. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth and its occlusion. This article presents a case report of Prosthodontic Management of a patient diagnosed with Dentinogenesis Imperfecta.

Keywords: Dentinigenous Imperfecta, Prosthodontics, Management.

INTRODUCTION

Dentinogenesis imperfecta (DI), also known as hereditary opalescent dentin, is a genetic disorder affecting the development of the dentin. This condition usually affects both primary as well as permanent dentition. The frequency of DI is believed to be about 1 in 6000 to 8000 people.[1] The DI condition usually causes tooth discoloration from an opalescent blue gray to dark yellow brown. Dueto the weakened dentinal structure, patients withthis condition generally presents with severely worn dentition, loss of tooth structure, missing teeth, and loss of occlusal vertical dimension.Also the teeth presents with short, constricted roots and pulpal obliteration due to dentin hypertrophy. Eventhough enamel in patients with DI appears normal in thickness and radiopacity, the underlying dentin which is defective causes detachment of enamel and thereby prone to severe wear.

Treatments for patients having DI are generally complicated and are difficult depending on the severity of enamel fracture, attrition, and pulpal involvement. Teeth affected with DI are mostly not prone to dental caries on comparison with normal teeth and this is considered to be a result of severe wear, which progresses faster than caries. Due to this, patients with DI often present with missing teeth and loss of vertical dimension. DI-affected teeth can be restored with composite resin veneers and bonded restorations Missing teeth due to wear and fracture often require extensive prosthodontic

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Received on: 31.10.2024

Accepted on: 07.12.2024

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rehabilitation and, in some cases, preprosthodontic orthodontic treatment. Pulpal obliteration or large pulp chamber (in DI type III) as well as the restorability of the remaining tooth structure can present challenges to endodontic therapy and restoration of these teeth after endodontic treatment.[2] Dental implant therapy can be a viable treatment option for nonrestorable DI affected teeth.

CASE REPORT

A female patient aged 45 years reported to the dental outpatient department with the complaints of discolored and disfigured teeth. (Fig. 1) She complained of the teeth being chipped off frequently and was concerned about her unesthetic, disfigured teeth, and felt very embarrassed to smile. (Fig. 3) She had abnormal dentition since childhood and had no history of any major illness. She did not report any history of such a dentition in the family, On clinical examination, the patient had generalized yellowish brown discoloration in the lower arch as well as the upper arch. (Fig. 2) The periodontal condition was found to be sound with satisfactory oral hygiene. A diagnosis of dentinogenesis imperfecta was made based on the radiographic finding and clinical examination.

The aim of the treatment was to remove the source of infection, restore function, esthetics and protection of posterior teeth from wear. The sources of infection to be removed by endodontic therapy, and this to be followed by full coverage crown for all the teeth to prevent further wearing of teeth.

TREATMENT PLAN

The treatment plan was explained to the patient and discussed in detail before commencing with the treatment. Risks, substitutes, and advantages of treatment were explained. Informed consent was obtained before treatment. In the control phase, It was planned to do scaling and root planning followed by treatment of decayed tooth.

In the definitive phase, it was planned to fabricate fixed prosthodontic restorations in the form of Crowns and Fixed partial dentures for both maxillary and mandibular arches.

TREATMENT

Control Phase

Initially, scaling and root planning was done for the patient, this was followed by endodontic treatment on 46. A diagnostic cast was fabricated for wax up purpose. The casts were mounted on a semi-adjustable articulator with the use of a face bow and centric record.

Definite Phase

Anterior teeth preparation was done from canine to canine in the maxillary and mandibular arch. (Fig. 4) Impressions were made with polyvinyl siloxane material. Provisional prosthesis were fabricated and luted, this wasfollwed by cementation with definitive prosthesis in the form metal ceramic crowns according to the anterior guidance. This was followed by preparation of the posterior teeth to receive metal ceramic crowns in 14, 15, 16, 17, 24, 34, 44, 45, 46, 47 and fixed dental prosthesis was given from 25 to 27 and 35 to 37. (Fig. 7) All the prosthesis were luted with GIC cement (Fig. 6) and post cementation instruction were given. (Fig. 5)

DISCUSSION

The term, 'Dentinogenesis imperfecta' was first introduced by Robert and Schour in 1939. Initially, Shields had divided dentinogenesis imperfecta into



Fig. 1: Discoloured Posterior tooth on Right side



Fig. 2: Discoloured Maxillary & Mandibular tooth



Fig. 3: Discoloured Posterior tooth on Left side



Fig. 4: Prepared Anterior tooth

3 types namely DI type I, II and III defects. Witkop named the types as dentinogenesis imperfecta, hereditary opalescent dentin, and brandywine isolate.[1] Later, this classification has been revised after the recent genetic studies which were done.

Type I is of the dental phenotype associated with osteogenesis imperfecta (OI), which is most commonly caused by genetic mutations of genes associated with collagen type I, COL1A1 and COL1A2.[3] Type II and type III are generally associated with mutations in the gene encoding dentin sialophosphoprotein.[4,5] Type III was denoted to describe a phenotype characterized by large pulp chambers that was first characterized in a triracial population from Maryland and Washington, DC.[6] Type III, also known as the Brandywine isolate,[6] is caused by mutations in the dentin sialophosphoprotein gene and thus represents a phenotypic variant of DI type II and not a separate entity. DI is a localized form of mesodermal dysplasia which is observed in the histo-differentiation,[8] Furthermore coarse and less number of dentinal tubules is seen in circumferential dentin with nonscalloped dentinoenamel junction. The area of void dentinal tubules also seen in dentin with less number of odontoblasts.[7]



Fig. 5: OPG with Crowns & FPD

In this article, the patient presented with Shields DI type II with worn dentition, multiple defective and unesthetic restorations and missing multiple teeth, but does not experience the common complication of loss of vertical dimension typically seen in severe cases of DI. We used fixed prosthesis to replace missing teeth and to help restore the occlusion. Fullmouth reconstruction was performed using fixed partial denture and crowns using metal ceramic restorations.



Fig. 6: Crown Placement on Maxillary & mandibular tooth



Fig. 7: Right Side view of Crown Placement in Both arches

Early diagnosis and treatment with restorative procedures like crowns or composite is crucial in

preventing significant tooth wear and preserving the vertical dimension in people with DI. Frequent folow up visits are necessary to monitor tooth wear and addressing any potential issues early on to maintain proper occlusion and aesthetics Treatments for patients with DI can be complex because of the loss of tooth structure and missing teeth. Full-arch splinted ceramometal crowns and FPDs were recommended by Bouvier *et al*.[9] Henke *et al.*[10] described a patient with DI who underwent full-mouth reconstruction using dental implants in the mandible while preserving natural teeth in the maxilla by restoring with full-arch splinted ceramometal crowns.

CONCLUSION

Dentinogenosis imperfecta presents a cumbersome task for the prosthodontist to achieve functional and esthetic restoration. Early diagnosis and treatment planning is crucial to obtain a favourable diagnosis.The present case uses metal ceramic crowns and Fixed partial denture to restore maxillary and mandibular arches to prevent tooth wear and loss of vertical dimention. Patients with DI may show a wide range of clinical features, therefore clinicians have to use their own judgment in selecting materials and treatments that are appropriate for the particular patient.

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