INDIAN JOURNAL OF SCIENCE AND TECHNOLOGY

CASE REPORT

Prosthodontic Management of Dentinogenesis Imperfecta - A Case Report

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OPEN ACCESS

Received: 05-07-2023
Accepted: 16-12-2023
Published: 13-01-2024

Citation: Kuriakose EM, Aradya A, Chandra KP (2024) Prosthodontic Management of Dentinogenesis Imperfecta - A Case Report. Indian Journal of Science and Technology 17(3): 228-235. https://doi.org/10.17485/IJST/v17i3.1660

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Funding: None

Competing Interests: None

Published By Indian Society for Education and Environment (iSee)

ISSN
Print: 0974-6846
Electronic: 0974-5645

Abstract

Objectives: Prosthodontic management of patients with Type II Dentinogenesis Imperfect, to provide a near to normal oral health for the individual, thereby improving their physical, mental and social well-being. **Methods:** Alginate impressions and border moulding was done. After a satisfactory try-in, the dentures were processed and delivered to the patient. Minor occlusal corrections were carried out during the insertion appointment and home care instructions were given to the patient. **Findings:** subject had normal primary dentition but the permanent dentition which replaced them fractured one after the other when it came into occlusion resulting in complete wearing out of his permanent teeth. Family history revealed that his sister also suffers the same condition with clinical manifestations like yellow-brown translucency on primary teeth. The child had a substantial loss in the vertical dimension of the face with a convex facial profile and retrognathic mandible and puckering of the chin. The lips were unsupported which gives a toothless appearance to him. Intra-oral examination revealed that the subject's occlusion is in a developing stage and whichever tooth that has come in contact with the opposing attrite to the gingival level. **Novelty:** The current patient has not been able to chew any food for the past 4-5 years since he lost his deciduous teeth. Such effects of DI can even affect the growth spurt and nutritional status of the individual. the patient has a severe attrition of teeth which has resulted in the loss of vertical dimension. So along with the restoration of mastication, the over-denture treatment also intends to increase the vertical height of occlusion to facilitate support of lips and overall growth of the facial-muscular entity. The clinical crown height of all the teeth was less and considering the patient's age over denture was delivered to the patient because of the economic constraints.

Keywords: Attrition; Dentinogenesis Imperfecta; Discolouration; Rehabilitation; Loss of vertical dimension

1 Introduction

Dentinogenesis Imperfecta (DI) as it was named by Robert & Schour in 1939is an autosomal dominant condition affecting both deciduous and permanent dentition. It was recognized by Barret in 1882and later was described as an enamel defect by Tabbot. DI was earlier termed as 'Hereditary opalescent dentin' by Skillen, Finn and Hodges which describes the brown translucent teeth that have an opalescent sheen. Affected teeth are grey to yellowish brown and have broad crowns with constriction of the cervical area resulting in a tulip shape. [1]

Aetiology of DI routes to dentin sialophosphoprotein in chromosome number. It constitutes 50% of the non-collagenous component of the dentin matrix. DI is inherited in an autosomal dominant fashion with high penetrance and low mutation rate. It is the most common genetic disease

Shield in 1973 had classified DI into 3 types:

- Type I DI is also associated with osteogenesis imperfecta.
- Type II DI is where osteogenesis imperfecta is not a feature.
- Type III DI is also known as Brandywine isolate which shows "shell teeth" with multiple pulpal exposure in primary dentition

Shield's classification was later revised based on the finding that dentinogenesis imperfecta and osteogenesis imperfecta are two different entities and the revised classification includes:

Dentinogenesis Imperfecta 1: DI without osteogenesis imperfecta

Dentinogenesis Imperfecta 2: Brandywine type dentinogenesis imperfecta which corresponds to type II DI of Shield's classification. [2]

Deficiency of dentin sialoprotein has been suggested as a causative factor by Zhang et al. in 2001. DI type II, III and dentin dysplasia are caused by mutations in dentin sialoprotein genes. [3]

Patients with DI may clinically present with amber, grey, yellow, brown, purple or bluish translucent discolouration of primary or permanent dentition. The severity of discolouration and extent of enamel fracture may be different among individuals belonging to the same family. In severe situations, the dentition may wear off to the gingival level. Tooth mobility, abscess or early loss may be also manifested. Sclerosed dentin with a hard glossy appearance often results in loss of sensitivity of teeth for the subject. [4]

On radiographs, the teeth have bulbous crowns with narrower roots and pulp chambers. Root canals may be smaller than normal or obliterated. Mac Dougall et al. (1998) stated that DG2 differ from DG1 by the presence of multiple pulp exposures, normal non-mineralized pulp chambers and canals, and classic "shell tooth" appearance. [5,6]

Histological appearances suggest normal enamel except for the peculiar shade which may be a manifestation of dentinal disorder. Dentin is composed of tubules with larger diameters, irregularly arranged with large areas of uncalcified matrix. Odontoblasts inclusion in the dentin may be present. The pulp chamber is often obliterated due to the continuous deposition of dentin. [7]

The teeth show a 60% increase in the water content and reduced inorganic contents compared to normal dentin. The density, x-ray absorption and hardness of the dentin are also low. The microhardness of dentin in DI close approximates that of cementum which explains the rapid attrition of teeth. [7]

Diagnosis of DI is predominantly done based on history, clinical examination and radiographic features. Molecular genetic testing can be used to confirm the diagnosis. As DI is an autosomal dominant disorder, there is a 50% chance that children born to affected parents be affected with DI. [7]

Differential diagnoses include hypo calcifiedamelogenesis imperfecta which results in exposure of underlying dentin or congenital erythropoietic porphyries or tetracycline stain which can cause intrinsic discoloration like in DI. Hypophosphatasia and immunological disorders like severe congenital neutropenia, Chediak-Hegashi Syndrome, histiocytosis X, Papillon-Lefevre syndrome and leucocyte adhesion deficiency syndrome can cause early teeth like DI-II. Vitamin D-dependent and Vitamin D-resistant rickets can show clinical and radiographical features of DI. [7]

The most noticeable oral feature in Osteogenesis Imperfecta [OI] patients was DI; in the primary dentition, yellow-brown type discolouration was more susceptible to attrition than the opalescent-grey type. For primary molars, stainless steel crowns are advised to avoid early tooth loss. [8] The prognosis of DI largely depends upon the age at which the diagnosis is given and the speed and quality of treatment provided to the subject.

Patients with DGI have teeth that are opalescent and brown, which gives them an unattractive appearance. They also have reduced masticatory abilities due to high attrition, which causes pain soon after teeth erupt. DGI Type I and II pulp chambers may initially be excessively wide, giving the appearance of shell teeth on radiographs, which later disappear due to hypertrophied dentine. [2] Since DGI Type I affects both dentitions, treatment necessitates many follow-up visits and involves the management of every tooth that erupts into the oral cavity, raising the overall expense of care. Frequent visits, the parents' socioeconomic

situation, knowing the child's psychology, and managing the child appropriately are a few of the significant difficulties in caring for young children that should not be neglected. The patient's aesthetic concerns must be taken into account because they affect their psychosocial behaviour. [9]

2 Methodology

A 14-year-old male patient walked into the outpatient clinic with the chief complaint of inability to chew food and slight pain in the upper right back tooth region.

A detailed history of the presenting illness was taken which revealed that the subject had normal primary dentition but the permanent dentition which replaced them fractured one after the other when it came into occlusion resulting in complete wearing of his permanent teeth (Figure 1). He never had any incidence of pain or sensitivity until recently he had pain in his maxillary right first molar tooth. The past medical history states that the boy was born after a full-term pregnancy but information about the milestones of development was not available since the child's parents were deceased. There has been no incidence of any other systemic illness or drug usage. Family history revealed that his sister also suffers the same condition with clinical manifestations like yellow-brown translucency on primary teeth.

Fig 1. Completely worn out permanent teeth

On extra oral examination, the child had a substantial loss in the vertical dimension of the face with a convex facial profile and retrognathic mandible and puckering of the chin (Figure 2). The lips were unsupported which gives a toothless appearance to him. Intra-oral examination revealed that the subject's occlusion is in a developing stage and whichever tooth that has come in contact with the opposing has attrited to the gingival level. The metal crown placed on the maxillary right first molar was tender on percussion and the mandibular right first molar was missing which was extracted two years back due to pain. All the deciduous teeth have exfoliated and the permanent counterparts have erupted. 38 and 48 were erupting when the patient was examined but were not in occlusion (Figure 3).

Fig 2. Extra Oral image showing loss of vertical dimension

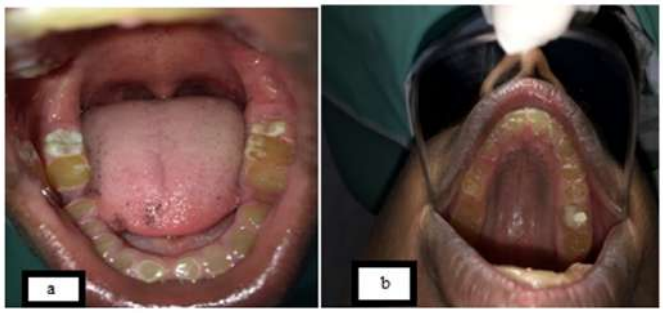

Fig 3. Intraoral view (a)-Mandiblar arch, (b)- Maxillary arch

3 Radiographic Examination

The orthopantomogram (OPG) (Figure 4) revealed bulbous crowns and marked cervical constriction of 17, 18, 27, 28, 37, 38, 47,48. A severely obliterated pulp chamber was found in 32, 34, 41, 42, 44, 45 and partial obliteration of pulp on remaining teeth. A supra eruption of 25 has been noted. A bony defect was detected between 35 and 36. Severe occlusal wearing was noted with 35, 36,47. Root completion was in progress of 18, 28, 38,48. The child's dental age determined was more than that of his chronological age.

Fig 4. Radiograph of the patient

4 Diagnosis and Treatment Plan

After correlating the clinical and radiographic findings along with positive family history, the child was diagnosed with Shield's DI type II. Considering the age of the child and the financial constraints of the family an over denture was planned to protect the remaining dentition and to enable chewing by the patient. The treatment plan was explained to the child's guardian and informed consent was obtained. Since the child was suffering from pain at 16, crown removal was done followed by root canal treatment before the fabrication of the denture. Alginate impressions were made of the upper and lower arch and study models were prepared using dental plaster. The cast was analyzed for soft tissue and hard tissue undercuts. Since most of the teeth were worn out and were at/below the gingival level, dental undercuts were minimal. Custom-made special trays were made using DPI cold cure acrylic resin to make a secondary impression on the study model with a double spacer on the occlusal surface to record it in minimal pressure. Border moulding was done using DPI Pinnacle tracing sticks and the secondary impression was made using zinc oxide eugenol impression paste (DPI Impression paste) (Figure 5). The final cast was poured using Type 3 dental stone (Goldstone).

DPI cold cure acrylic resin was used to fabricate the denture base and the labial flange of the upper denture base was removed to relieve the undercut which will hinder the placement. The occlusal rims were fabricated and since the vertical dimension of the subject was collapsed, a vertical dimension was recorded and teeth selection was done. The maxilla mandibular record was

Fig 5. Secondary impression of maxillary and mandibular arch

transferred onto the 3-point articular and mounted using plaster of Paris. The teeth arrangement was done on the articulator and try-in was done on the patient. (Figure 6)

Fig 6. Try in procedure

After a satisfactory try-in, the dentures were processed and delivered to the patient (Figure 7). Minor occlusal corrections were carried out during the insertion appointment and home care instructions were given to the patient. The weight of the patient was checked at the denture insertion appointment and was recorded as 46kg. The patient was recalled on the next day for a review visit over 24 hours of denture wearing and later after one month. The patient reported that he was able to chew his food properly with the denture. The weight recorded at that visit was 47kg.

Patient informed consent was taken with the reference number of 9556/20.

Fig 7. Insertion of dentures

Fig 8. Post Insertion side Profile

5 Results and Discussion

The major components of the preventative strategy for individuals with DI include strict oral hygiene and dietary guidelines, as well as topical fluoride varnish application. It is essential to keep close/regular supervision, as well as follow-up visits, for maintaining good oral health.

In the early stages, between the ages of 18 and 20 months, and in the later stages, between the ages of 28 and 30 months, severe DI can be initially treated. Other dental professionals (such as prosthodontists, orthodontists, periodontists, and maxillofacial surgeons) are frequently needed for interdisciplinary therapy later on during mixed dentition.

To restore function and aesthetics, dental therapy may be divided into three phases: (i) treatment of primary teeth; (ii) provisional treatment of permanent teeth; and (iii) definitive treatment of permanent teeth. Overdentures restore aesthetics, vertical dimension and function. Additionally, it is feasible to produce a comparatively stable occlusion, which increases the patient's tolerance for subsequent procedures for treating worn teeth. However, plaque retention spaces may cause hygiene issues.[(10)]

Mastication is considered one of the most important functions of the stomatognathic system as it facilitates the digestive procedure. The absence of teeth primarily affects the food intake by the individual more than the appearance and growth of the oro-maxillary complex. The effect of DI on young individuals can range from physical, mental and psychological.[(10)] The current patient has not been able to chew any food for the past 4-5 years since he lost his deciduous teeth. Such effects of DI can even affect the growth spurt and nutritional status of the individual.

The present individual reports having 'normal' teeth during his young age and the new set of teeth which erupted later were all worn out eventually. The patient was not able to get the condition diagnosed earlier nor could he avail any treatment because of his economic backwardness and family conditions. Treatment strategies for DI are selected based on the age of the patient, the severity of the problem and presenting complaint of the patient. Hence, it is very important to educate such individuals by conducting rural-based oral health camps and also by other mass media.

In this case, the patient had lost all his primary teeth and permanent teeth which are already in occlusion had already worn out severely and the remaining permanents are not yet in occlusion. So overdenture treatment was provided for the patient taking into consideration of his age and economic factors. Literature has suggested that DI of permanent teeth can be managed by using:

- Porcelain veneers or porcelain crowns
- Full mouth rehabilitation using crowns and partial dentures
- Overdentures in case of severe tooth wear.
- Dental implants after 18 years and growth is completed.[(11)]

Studies have suggested the use of overdenture as a treatment option in severely worn-out permanent dentition. They also emphasized minimal tooth preparation until the child reaches childhood. Researchers have suggested the choice of replacement

of teeth till the child completes 18 years is complete dentures and dental implants can be considered once the growth is completed.[12,13]

In the present case, the patient has a severe attrition of teeth which has resulted in loss of vertical dimension. So along with the restoration of mastication, over denture treatment also intend to increase the vertical height of occlusion to facilitate support of lips and overall growth of the facial-muscular entity. Studies have observed that patients requiring a definitive prosthesis for oral rehabilitation will tolerate a moderate alteration to the vertical dimension of occlusion.[14,15]

They also suggested that since the definitive treatment is time-consuming and expensive, it is preferable to use it as a prosthesis that does not permanently change dentition during the usage period. In such cases, overlay denture or overdenture is an effective treatment option. Alvitez T. D et al.[12] suggested that an increase in the vertical dimension of occlusion in children shows no or less than 5% discomfort among the subjects if a stable occlusion is given. In the present case, the subject was able to chew on food and was comfortable with the change in the vertical dimension of the face, but a lip incompatibility was noted on the day of denture delivery which the patient was not concerned about.

A normal acrylic denture was delivered to the patient because of economic constraints. The presence of the remaining tooth structure provided commendable stability and retention to the denture. Studies have suggested the usage of silanized glass fibre to prevent midline fracture due to flexural fatigue and impact force.[16]

Overdentures retained and supported by abutment teeth or dental implants are another restorative option for older individuals or those with severely deteriorated dentitions. DSD can be a key component in producing an acceptable outcome in terms of aesthetic design features. A multidisciplinary treatment plan is also quite helpful in restoring a patient's appearance who has DGI-I.[2]

The weight of the patient was noted on the day of denture delivery to assess its improvement. The patient on his recall after a month reported to be satisfied with the denture and was able to chew his food. The weight assessed at the recall appointment showed an increase of one kilogram of weight and the patient was smiling with confidence. The importance of maintenance of oral hygiene and denture maintenance was repeated during the recall visit. The lip compatibility had improved on the recall visit and the patient was very accustomed to the prosthesis.

These teeth's crowns look bulbous because there seems to be cervical constriction. As part of this treatment, the anterior teeth's aesthetic is restored, the occlusal vertical dimension is maintained, and structural protection is used to prevent cavities from developing and wear from abrasion. Additionally, a multidisciplinary approach, aesthetics, and differential diagnoses should be taken into account during the therapy procedure.[2]

Alrashdi M et al. highlighted in their study including siblings that both children were put on a regular recall schedule that included an oral examination (every 6 months), dental prophylaxis, oral hygiene training, and, in the case of the first patient, denture adjustment, if necessary. This was done because the child's growth might require relining or remaking the denture.[14] When development is complete and an implant can be inserted, follow-up treatment should ideally continue. Another choice regarding the overlay denture's fabrication was to include an expansion.[14] In contrast to fixed applicants, the use of Clear aligner Treatment (CAT) in patients with Amelogenesis Imperfecta [AI] and Dentinogenesis Imperfecta [DI] is advantageous. However, its administration should be done with caution; due to the decreased highest point of the crown and absence of undercuts. As a result, aesthetics should be checked and improved whenever possible to encourage patient compliance. The cost of CAT is higher than that of fixed appliance therapy. Additionally, creating a plan for fixed appliance therapy takes the orthodontist less time than designing a strategy for clear aligner treatment.[13]

Authors have recently published a clinical report showing how a DI-II patient was successfully treated using a variety of digital techniques, such as DSD, the ARCUS digmaaxiograph, and CAD/CAM. The successful treatment of this patient with DI-II also required an interdisciplinary prosthodontic/periodontic approach.[16]

6 Conclusion

Lack of knowledge and awareness among the people of the treatment of dentinogenesis imperfecta can severely affect the prognosis of the overall treatment outcome. Early recognition and diagnosis of the condition enable the practitioner to provide a near to normal oral health for the subjects, thereby improving their physical, mental and social well-being. This case report also calls for improving awareness among the general community towards such conditions through rural health camps and mass awareness through media because "catching them young" can improve the prognosis and help such individuals to lead near-to-normal life.

A favourable prognosis for DGI depends on early diagnosis and therapy. The preservation of remaining teeth while restoring their function and appearance is of the highest importance. In the late stages of the primary dentition, prosthodontic rehabilitation of a child with severe DGI considerably improves function, and aesthetics, and shows to be a considerable psychological boost to the patient's well-being. We recognize the value of further research in this area.[14]

As a result, we strongly advise establishing and planning such research approaches to use the suggested results derived from case reports and evaluate materials and methods for teeth with severe wear from the modern concept of Evidence-Based Dentistry.

Preventing future tooth fractures and restoring oral function are the key concerns for DI patients. To avoid peer pressure, aesthetic care becomes more crucial in young children. A less expensive approach that is easier to cure, modify, and retreat in emergencies. It offers greater flexibility in formulating treatment strategies.

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