Restorative Management of Dentinogenesis Imperfecta in an adult - A case report

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Abstract

Dentinogenesis Imperfecta is a localized mesodermal dysplasia affecting both the primary and permanent dentition. First described in the late 19th century, it is characterized by discolored and translucent teeth ranging from grey to brownish-blue or amber. Dentinogenesis Imperfecta, type II (DGI-2) is rare and it is a severe form of the condition. Radiographically, the crowns of the teeth are bulbous with marked cervical constrictions, and the pulp chambers become obliterated over a period of time. Sensori-neural hearing loss has also been reported in some patients.

This case report presents the restorative management of DGI-2 in a 44year old clergy woman whose primary complaints were poor aesthetics and lack of social acceptance. The case was managed with consideration of the patient's limited financial resources at the Lagos State University Teaching Hospital, Ikeja (LASUTH). The objective of the management of this condition was to open the bite of the patient using maxillary posterior bite plane followed by intermediate restoration of worn teeth using composite resin for the newly established occlusion. The patient used the restorations for a while before definitive restorations, using ceramo-metal crowns as well as over dentures, were carried out.

The restorative treatments corrected the patient´s vertical dimension of occlusion with resultant acceptable aesthetics and function. The patient was satisfied with maxillary anterior ceramo-metal crowns and a posterior removable over denture as well as a complete mandibular over denture.

Key words: Dentinogenesis imperfecta; Restorative treatment; Over-denture.

Introduction

Dentinogenesis imperfecta (DGI) is a hereditary dentin developmental disorder that affects both primary and permanent dentitions[1]. It is inherited in a simple autosomal dominant mode with high penetrance and a low mutation rate[2]. It is the most common group of hereditary dentine defects with a reported incidence of 1:6000-1:8000[3, 4]. It has an equal gender distribution and it is commoner in whites than in blacks[5]. There are 3 types of DGI[6]. Type I is associated with osteogenesis imperfecta and may be associated with midfacial hypoplasia, class III malocclusion, and posterior crossbite. Type II is the classical heredity opalescent dentin with yellow, amberbrown or bluish grey translucent teeth[6, 7]. Takagi and Sasaki suggested that the dentin in DGI type II is deficient in the phosphorous ion, which is important in the early stage of odontoblastic differentiation and its mineralization[8]. Pulpal obliteration is the radiographic feature of both the deciduous and permanent dentitions. Type III is the Brandywine form associated with a large population in the city of Brandywine, MD.

Management of DGI involves taking a good history to determine the involvement of primary dentition and a detailed family history with family pedigree construction. This includes history of exposure to chemicals, fluoride, drugs and trauma. History of bone fracture with minimal trauma, joint hypermobility, hearing loss should also be taken. Clinical examination to determine teeth colour, wearing of teeth, as well as investigations such as

radiographs and pulp sensibility tests should be done. Molecular genetic diagnosis could also be done. Restorative treatment focuses on retaining the remaining tooth structure and protecting the affected dentin from caries, attrition, abrasion and erosion. Prosthodontic treatment plan includes consideration of function, aesthetics and vertical dimension.

Toothwear (TW), a feature of DGI, can be managed by conventional restorative techniques (mechanical tooth preparation for retention and resistance form) or adhesive additive approach (adhesive retained restorations)[9]. Current concepts embrace the latter as the first line of management. Thus serving as a medium or intermediate restoration which is eventually replaced by conventional techniques following patient's tolerance and adaptability to the newly established occlusal scheme[9,10]. Adhesive additive approach involves restoring TW with direct or indirect composite resin as medium or intermediate definitive management of anterior teeth. Bevenius et al[11] were pioneers of the use of composite resin using acid etch technique. Hemmings et al[12] reported that hybrid resins offer more rigidity than microfil resin; and they recommended direct composite resin as a treatment option for localized anterior TW.

However, Bartlett et al[13] reported that the use of direct and indirect composite resin on posterior teeth will result to failure most especially in patients with parafunctional tooth grinding / clenching habit. Subsequently, Akar and Dundar[14] elucidated that glass fibre reinforced composite resin, with better wear resistance and minimal shrinkage, are alternative treatment mordality for localized anterior TW. Likewise, Artglass which is a ceromer was introduced by Gow and Hemmings[15] as an indirect composite resin palatal veneer serving as a fixed Dahl's appliance[4].

Aristidis and Dimitis[16] confirmed 90% success rate of porcelain laminate veneers for conservative treatment of anterior TW. Nohl et al[17] also reported 89% success rate for the use of cast alloy restorations in maxillary anteriors affected by acid. This was corroborated by Mehta et al[4] that cast adhesive alloys such as gold type III and Nickel-Chromium (Ni-Cr) are used for fixed metallic restorations in the form of “gold hats” or “bonnets” for posterior adhesive onlays or as anterior palatal veneers known as “palatal shims”[3,4]. Yab[18] recommended resin bonded onlays where there is plan to increase the Vertical Dimension. In such a situation, adhesive ceramic veneers are used following space creation by Dahl's principle with an initial short term composite resin restoration. Ceramic onlays and dentine bonded crowns are recommended for posterior teeth[4]. Oh et al[19] suggested the post treatment use of occlusal splint as nocturnal use for protection of all restorations from associated parafunctional habits which may result to premature degradation of these restorations.

The objective of the management of this condition was to open the bite of the patient using maxillary posterior bite plane followed by intermediate restoration of worn teeth using composite resin for the newly established occlusion. The patient used the restorations for a while before definitive restorations, using ceramo-metal crowns as well as over dentures, were carried out. There has been no record of the treatment of this type of condition in this establishment; hence, we are presenting a case of DGI type II in a 44 year old clergy woman whose primary complaints were poor aesthetics and lack of social acceptance.

Case report

A 44 year old clergy woman presented at the Lagos State University Teaching Hospital, Ikeja (LASUTH) with complaints of poor aesthetics, unsatisfactory social acceptance and difficulty with chewing **(Figures 1 and 2)**.

Figure 1: Pre-op clinical photograph

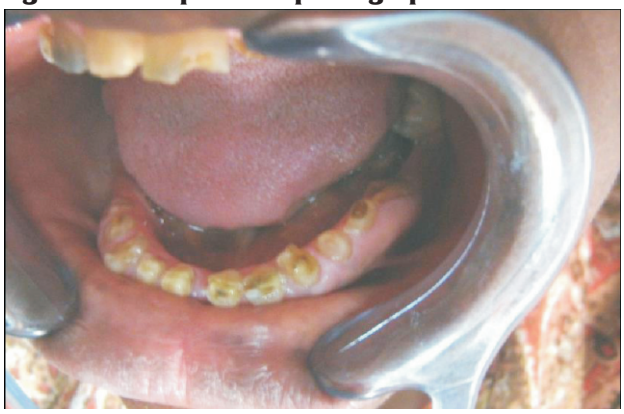

Figure 2: Pre-op clinical photograph

There was a positive history of discoloured and brittle teeth from childhood. There was no significant history of childhood infections, excessive exposure to fluoride or medication usage or any underlying medical condition. Patient had previous dental visits for scaling/polishing and extractions which were uneventful. There was no history of any significant childhood illness neither trauma to the teeth nor of bone fractures with minimal trauma to any part of the body. There was also no history of hearing impairment. A detailed family history revealed a positive history of similar condition in her grandmother, mother and sister. Two of her children having similar dental problems with variable severity.

On examination, the characteristic pale blue sclera consistent with osteogenesis imperfecta was absent. There was no facial asymmetry while the lips were competent. The rest vertical dimension was 64mm while the occlusal vertical dimension was 60mm. Intra-oral examination revealed abnormalities of dentition consistent with findings in DGI-2 in all the teeth present. All oral soft tissues were clinically healthy while the oral hygiene was fair (OHI-S = 1.8). There was generalised brownish yellow discolouration of the teeth. There was also severe attrition with labial, buccal and occlusal wear (Figures 1 and 2) of most of the teeth with a resultant decreased vertical dimension but no associated tooth sensitivity. Patient had a deep bite resulting in a bite of convenience. There were no carious teeth and all the teeth had Grade 2 mobility. Tooth 11 had Ellis Class 2 Ellis fracture.

Teeth present:	7 6 5 4 3 2 1	1 2 3 4 5
	5 4 3 2 1	1 2 3 4 5 6 8

Full mouth periapical radiographs and Orthopanthomogram (OPG) revealed periapical radioluscency around the upper right canine. There was also severe horizontal bone loss around retained roots of the upper right first premolar and first molar. The upper right lateral incisor and canine were grossly broken down and there was gross attrition of the coronal surfaces of the upper left lateral incisor, canine, first and second premolar. There was also a slight widening of the periodontal ligament space around lower left central and lateral incisors. There was generalized obliteration of all root canals. Pulp vitality tests (cold test using ethyl chloride) was negative for all teeth. A clinical diagnosis of DGI-2 was made.

A problem list of poor aesthetics, inefficient mastication and reduced vertical dimension associated with deep bite was made and the treatment objectives were to recover the lost vertical dimension, improve aesthetics and social acceptance. Treatment plan was drawn up which involved taking clinical photographs and making study models. Oral health education and dietary counseling as well as scaling and polishing were also included in the plan. This was followed by the definitive restorative treatment.

Three stages were established in the definitive restorative treatment. The first stage was preparatory stage which included oral health education, dietary counseling and oral hygiene maintenance programme. She was educated and motivated in oral hygiene maintenance. Scaling and polishing was done. Flouride prophylaxis was carried out using topical fluoride varnish. The second stage was the surgical stage in which grossly worn down upper right canine, first premolar, first molar and the lower left third molar were extracted while deep scaling and root planning of lower left central and lateral incisors was done. The third stage was the restorative stage involving full mouth restoration. This was commenced by opening up the bite of the patient through increase in the occlusal vertical dimension using the maxillary posterior bite plane (MPBP). The thickness of which was initially kept at 1mm and thereafter gradually increased in increments of 1mm up to 3 mm, by adding self cure clear acrylic over already processed heat cure acrylic.

Subsequently the maxillary and mandibular posterior removable over-dentures were provided following rounding up of root stumps of the upper left premolars; the lower left incisors, canine premolars and first molar; and the lower right incisors, canine and premolars. (Figures 4 and 5). The patient was satisfied with the treatment outcome (Figure 6).

This gradually opened up the bite. The (MPBP) was worn by the patient for a period of three months at an established occlusal vertical dimension which she was able to tolerate biologically without any discomfort at the right and left temporomandibular joints. During this period the patient was reviewed regularly on a monthly basis. Once the opening of the bite and increase of the vertical dimension were achieved, there was the provision of fixed and removable prostheses in order to achieve full restoration of the mouth.

The fixed prosthesis comprised of individual anterior ceramo-metal (porcelain fused to metal-PFM) crowns fabricated for the upper right central incisor, lateral incisor and first premolar and the upper left central incisor, lateral incisor and canine. **(Figures 3 and 5)**.

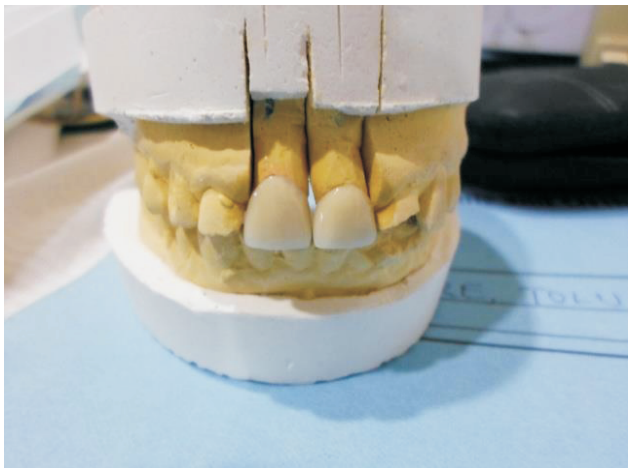

Figure 3: Porcelain fused to metal crowns for 11 and 21

Figure 4: Finished over-dentures.

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Figure 5: Appearance after treatment

Figure 6: Patient satisfied with treatment

She was recalled quarterly for check-up for one year and subsequently every 6 months up till date. At each recall visit, the patient was asked if she had a fresh complaint after which she was thoroughly examined clinically. Extra-orally, she was observed mainly for facial asymmetry and temporomandibular joints anomalies such as tenderness and clicking sounds. Intraorally she was examined to check the integrity of the oral hygiene, oral mucosa, the oral faces of the retained root stumps for evidence of root caries. The porcelain fused to metal crowns were also checked for failures such as porcelain cracks, fracture and compromised retention. Orthopanthomograph and periapical radiographs were taken to monitor the health of the supporting alveolar bones and roots for any pathology. Once all was well, oral hygiene instructions were re-emphasized. Topical fluoride varnish was applied on each root stump. The upper and lower removable over-dentures were also examined for cleanliness and for fit.

Discussion

The rehabilitation of any case of dentinogenesis imperfecta is a challenging venture for the dental practitioner because this full mouth restoration is usually complex and requires a multidisciplinary effort of several aspects of dentistry. First, there should be preventive measures to improve oral hygiene of the patient. This patient was educated and motivated on her role in oral hygiene maintenance programme. In DGI, the histologic structure of the dentin appears relatively normal but the number of the dentinal tubules is decreased. The characteristic scalloping at the dentinoenamel junction is diminished or absent. This scalloping is supposed to provide mechanical interlocking between dental hard tissues. Once this is missing the enamel is easily detached resulting in teeth with exposed dentin which exhibits severe abrasion or fracture. The

exposed dentine takes up stains from food and beverages and could also be prone to sensitivity[20,21,22,23,24,25]. The patient in this case study had no sensitivity. This could be due to presence of irregular sparse dentinal tubules or complete absence of dentinal tubules and obliterated root canals.

Flouride was used as a prophylactic measure to increase resistance of root stumps to caries. The scaling and polishing procedure that was done prior to restorative treatment brought about a tremendous improvement in the periodontal status of the teeth which became firmer in spite of their grade 2 mobility at the onset of management. This patient had to be motivated and counselled because management of DGI is usually complex and involves several visits to the dentist with a great deal of financial implication. This financial implication, of multiple dental visits for over a period of more than one and a half year, was significant to this patient. Endodontic treatment of teeth with DGI is usually avoided because in most cases the root canal are narrow or absent[5]. In this study, the extracted teeth were diagnostically found, through radiographic and clinical evaluations, to be periodontally compromised, had obliterated root canals and thus resulting in poor prognosis.

The full mouth rehabilitation of this case study was preceded by the gradual opening of patient's bite by increasing the vertical dimension of her occlusion. There was an initial determination of the amount of vertical dimension that could be increased without causing discomfort or affecting the balance of her temporomandibular joint (TMJ). This was achieved by determining the existing freeway space (FWS), the closest speaking space and vertical dimension of occlusion. The patient's FWS was in the range of 3-5mm.The initial thickness of the maxillary posterior bite plane (MPBP) was 1mm and was gradually increased to 3mm by the addition of self cure clear acrylic over already processed heat cured acrylic. Following the use of the (MPBP), the vertical dimension of occlusion was increased. This was biologically acceptable for this patient.

Following this bite opening, the standing upper anterior teeth were clinically and radiographically re-assessed and it was decided that the upper anterior crowns could be prepared without surgical crown lengthening because they had adequate crown-root ratio. This was in agreement with the fact that conditions of DGI often times, present with teeth that possess short roots thereby contravening the use of crown lengthening procedure[26].

There are several definitive treatment options for full rehabilitation of conditions of DGI such as complete coverage all-ceramic crowns, ceramo-metal crowns (Porcelain Fused to Metal-PFM), removable overdentures or implants for permanent dentition. Overdenture is utilized in cases of extreme abrasion or in older patients[5]. This particular patient was treated with a combination of individual ceramo-metal (PFM) crowns on the anterior teeth of the upper jaw where the crown-root ratio was not unbearably compromised by tooth wear hence there was adequate crown length for the crown preparation. The remaining posterior teeth on the upper jaw were restored with removable partial overdenture. The lower jaw was restored with complete removable overdenture. Other factors that were responsible for this choice of management included the limited financial status of the patient as well as the limited technological equipment and materials at this centre. The patient was quite satisfied with the outcome of the final treatment.

Studies have shown that in the more advanced environment, patients with DGI reported earlier as recorded by Helenia,[26] Francisco[27] and their co-workers. They treated 13year old boy and 11year old girl respectively. All ceramic crowns were used for full mouth restoration in the 13year old boy. The 11year old girl got her mouth rehabilitated with a combination of ceramo-metal (PFM) crowns in the anterior segments of her upper and lower jaws while she had removable skeletal prosthesis for the posterior segments. In the present study as well as with previous studies, it was apparent that complete coverage of crowns are usually the preferred restoration for the patients with DGI because such restorations protect the dental and alveolar bone tissues from further destruction.

All ceramic crown as an option, is said to be attractive due to combination of lower surface hardness, accuracy of fit, biocompatibility, and aesthetics[28]. In addition they can be cemented adhesively with resin cements and bonding agents providing increased dentin-to-porcelain bond strength. This gives a significant advantage when short teeth or abutments with irregular shape that cannot offer adequate retention are restored[29,30]. Likewise, ceramo-metal (PFM) crowns also create adequate aesthetics.[(27)] However, their use in opposition to unrestored teeth suffering from DGI should be avoided to prevent further abrasion[28]. Ceramo-metal (PFM) crowns are preferable in condition of heavy or deep bite and in people who chew a lot of hard foodstuffs. The patient's restorations had been in clinical use for over a period of 2 years without any functional problems and complaint from her.

Conclusion

Dentinogesis imperfecta type II (DGI-II) is a localized form of mesodermal dysplasia of the dentin affecting both the primary and permanent dentitions. As this condition is inherited in an autosomal dominant fashion, there is a 50% chance that a child born of an affected parent will also be affected. The management of this condition is challenging and requires a multidisciplinary efforts of different specialties in dentistry. However, early diagnosis of this condition is pertinent so as to prevent deterioration of teeth and its resultant negative effect on function, aesthetics and social acceptance. Aesthetically acceptable appearance is one of the primary requisite for social acceptance in a civilized world and a pleasing dentition is a significant contributor to aesthetics of the human face.

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