A Minimally Invasive Restorative Approach of Dentinogenesis Imperfecta: A Case Report

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Abstract

Dentinogenesis Imperfecta (DI), is a genetic disorder that can greatly impact the oral condition both in the deciduous and permanent dentition. In the past, treatment of DI patients has involved extensive prosthodontic treatment, which even in young patients could range from full coverage crowns to implant retained bridges. The aim of this article is to introduce a novel and conservative approach to restorative treatment of such patients using modern adhesion technology. The results obtained indicate the possibility of successfully treating such patients without having to undergo invasive treatment.

Keywords: Dentinogenesis Imperfecta; Pocelain Veneers; Conservative Restorative Treatment

Introduction

Dentinogenesis Imperfecta (DI) is a genetic condition which can affect both deciduous and permanent dentitions. It is an autosomal dominant disorder of tooth development characterized by the presence of opalescent or translucent dentin, resulting in brownish to blueish coloured teeth. DI was first reported in the late 1800s and first published case was in the 1970s [1]. It affects 1:68000 patients in the population, making it the most common genetic dental condition. There are also known associations with other conditions like Osteogenesis Imperfecta (OI) and Dentin Dysplasia (DD) [2].

The first classification introduced to describe DI was described as the Shields classification [2]. More recently, through molecular analysis and because of the different possible phenotype mutations, a modified and more extensive Mendelian Inheritance in Man (MIM) classification [3] has also been introduced, though there is still a lack of genetic information to propose a new and comprehensive classification to describe this complex genetic condition. Therefore based on what is known, the Shields classification remains the most popular and describes three types.

Patients with type I DI also have osteogenesis imperfecta. In both permanent and deciduous dentitions teeth have an amber and translucent colour and show marked attrition. Radiographically, the teeth have short, constricted roots and dentine hypertrophy leading to pulp obliteration either before or just after eruption. With some patients signs may range from teeth showing total pulp obliteration while in others the dentine appears normal [4].

Type I and II DI have similar dental features, but in type II osteogenesis imperfecta is not a feature. Furthermore normal teeth are never found in type II DI. Sensorineural hearing loss has also been reported as a rare feature of the condition [5].

There is a form of DI found in a tri-racial population from Maryland and Washington DC known as the Brandywine isolate. Though they may vary, this type has similar features to those seen in type I and II DI. Deciduous teeth show multiple pulp exposures and radiographically, they often manifest “shell” teeth which are characterised as hollow due to hypotrophy of the dentine [6].

Because of the varying nature of the dental implications, treatment of child and adult patients, can diverge from bleaching in less se-
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vere cases [7], to more comprehensive and invasive restorative therapy, such as full contoured crowns and implant therapy [8, 9, 10, 11, 12, 13]. Though, because of the generalised nature and often severe tooth substance destruction, conventional treatment of such patients involved extensive restorative treatment, which lead to further removal of tooth structure.

Case Report General Anamnesis

The 22 year old female patient presents to the surgery for consultation asking to improve the appearance of her teeth (Figure 1) and to get a second opinion regarding a tooth which the previous dentist started to treat as a small carious lesion, but ended up removing an extensive amount of tooth structure, that it had to be restored with a temporary crown. Her medical status is clear; she doesn’t take medications and has no allergies.

![Figure 1: Initial smile](image1)

Dental Anamnesis

Clinically we observed a generalised greyish to brownish colour of all her natural teeth (Figure 2), and further investigation revealed that this condition was present in other older members of her family on her father’s side. Closer examination of her natural teeth showed a moderate amount of occlusal and incisal enamel wear (Figure 3a, 3b) for her young age, diagnosed as an anterior clinical erosive (ACE) class III [14]. The extra oral lateral view (Figure 4) may even suggest that even at this stage some reduction of vertical dimension of occlusion (VDO) may have taken place due to the moderate amount of wear.

![Figure 2: Initial intraoral frontal view](image2)

Radiographic examination (Figure 5) revealed she has numerous teeth restored with crowns, some of which are implant supported. Periapical radiographs (Figure 6) showed generalised bulbous crowns and obliterated pulp chamber and canals of all her remaining teeth.

A differential diagnosis based purely on the clinical appearance of her teeth may have been a severe case of tetracycline staining, however both the clinical and radiographic evidence, and absence of other systemic effects caused by OI, she was diagnosed with type II DI.

**Figure 3a:** Initial intraoral occlusal view of upper teeth.

**Figure 3b:** Initial intraoral view of power teeth.

**Figure 4:** Initial extraoral lateral view with reduction in vertical dimension of occlusion.
Treatment Options

The goals of the therapy were to enhance dental and overall smile aesthetics, by improving tooth colour, restoring normal tooth height to width ratio of upper and lower anteriors and protecting her dentition from further wear. Regardless of the material chosen, the scope of the therapy was to achieve both functional and aesthetic goals, using as a minimally invasive alternative as possible. The possible options for the anterior teeth ranged from labial direct composite restorations, to labial and palatal indirect composite restorations, to porcelain veneers and indirect palatal composite veneers. Whereas for the posterior teeth the options were either composite resin or lithium disilicate occlusal indirect restorations.

The advantages and disadvantages of each option were discussed with the patient. Though initially direct composite restorations were given as a treatment alternative, a trial restoration was placed and was excluded as an option due to the inability of the composite restorations to adequately mask the darkened appearance of her teeth. Therefore the patient opted for porcelain veneers for both the upper anteriors and lower anteriors. In the posterior teeth Ultimate composite onlays were chosen. To match the restorative materials between the upper and lower arches lithium disilicate CADCAM veneers were chosen.

Treatment Carried Out

Due to the difficult nature of the case, before treatment was begun, several stages of aesthetic trials were carried out. A direct mock up was done to begin to study possible tooth length and form (Figure 7). This was followed by an indirect laboratory made mockup (Figure 8) using a diagnostic wax up of both arches. To try and improve the colour of the teeth vital bleaching was done (Figure 9a, 9b), though

this resulted in only temporary improvement of her colour. Due to the dark pigmented dentine, indirect feldspathic porcelain veneers or direct composite resin veneers would not adequately improve her colour. It was decided to use CADCAM made lithium disilicate copings layered with enamel porcelain. Therefore the next phase consisted in the colour trials of the lithium disilicate copings of the labial veneers. A single coping was made for the 11 in three different lithium disilicate shades: low translucency (LT) bleach, LT bleach 1 (LTB1) and medium opacity (MO). Each of these copings was trial cemented over the patient’s tooth with glycerol, A3 Variolink cement and D2 composite and the results were photographed (Table 1), noted, and LT B1 coping was chosen as the most suited coping in terms of masking capacity and naturalness of colour.

Figure 7: Direct mockup.

Figure 8: Indirect mockup.

Figure 9a: Preoperative bleaching.
In the first operative session the upper (Figure 10a) and lower onlay and lower veneer preparations (Figure 10b) and impressions were taken. Both arches were temporised using the lower full arch and upper posterior diagnostic wax-up made silicone key, immediately increasing the VDO and bringing into occlusion the posterior teeth, whilst leaving her with an anterior open bite (Figure 10c). At the next appointment the lower veneers (Figure 10d) and onlays were cemented. Next the upper onlays and palatal disilicate veneers were cemented and upper labial veneer preparations (Figure 10e) and impression were taken. Finally the labial veneers were cemented (Figure 10f). The final restorations were CADCAM manufactured Ultimate composite onlays and CADCAM manufactured lithium disilicate palatal veneers, and lithium disilicate copings stratified with porcelain for the labial upper and lower veneers.

Finally the 41 temporary crown was removed and the tooth was assessed. This tooth was severely reduced due by the previous dentist. Though the patient was advised of the questionable prognosis of this tooth, a direct composite core was built up on the tooth since no root canal was able to be carried out due to the obliterated canal. The final crown preparation was made and impression taken for a lithium disilicate crown was taken and cemented, matching the colour to the cemented veneers (Figure 10g). The patient was warned that the prognosis of this tooth and its restoration remained questionable due to the severe loss of tooth structure and poor adhesion of the composite onto the dentine. If and when this restoration would fail an implant retained crown would be placed. She was issued an upper occlusal splint and was brought back for a six monthly recall (Figure 11a, 11b).
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**Figure 10a:** Intraoral occlusal view of posterior onlay preparation.

**Figure 10b:** Lower veneer preparation.

**Figure 10c:** Temporisation of posterior onlays and lower veneers with resulting anterior open bite.

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Figure 10d: Final porcelain lower veneers and temporary crown 41.

Figure 10e: Upper veneer preparation.

Figure 10f: Final upper porcelain veneers.

Figure 10g: Final lithium disilicate crown on 41.

Discussion

It is generally accepted that the tooth’s hue is mainly given by the colour and properties of the dentine. The varying thickness of enamel layer acts to reduce the intensity of the tooth’s color, therefore not changing the hue, but varying the chroma or degree of saturation based on its thickness [15, 16]. That is why intrinsic tooth staining such as dentine defects, affect the tooth’s appearance and are more complex to resolve than intrinsic enamel defects or extrinsic staining.

The greyish to brownish opalescent discolouration observed in DI patients is due to the abnormal dentine colour, though the enamel has its close to normal structural and optical characteristics [5]. This explains why at the cervical third, where the enamel layer is at its thinnest, the discolouration is more pronounced. Under SEM, the dentine in DI patients shows many large and irregularly organised dentinal tubules, often missing the odontoblastic processes, and irregularly organised and distributed collagen bundles and fibers of abnormal size [17]. In addition, the dentino-enamel junction (DEJ) is significantly affected by DI, there is a marked reduction in the bond be-

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tween enamel and dentine [18]. Though dentine is the more affected of the two, some enamel abnormalities can be observed in scanning electron microscopy (SEM), such as irregular surfaces which exhibit cracks and crevice formation, evident signs of a weakened structure in sagittal sections [19]. Some authors [20] have recently concluded that the DEJ complex plays and important role in blocking enamel cracks propagation by deflecting the crack paths along it. This is consistent with the clinical findings of patients with DI who exhibit both significantly affected DEJ and multiple fracture lines. Though the exact reason why the characteristic loss of enamel of DI patients occurs is still not known, possible reasons may be due to the defect in dentine causing the mantle dentine to shear from the circumpulpal dentine [21], others believe the combination between low density apatite crystals of the dentine, abnormal collagen fiber orientation and enamel irregularities may be a potential cause, and may occur mainly at the DEJ [22].

More recent literature which examined the physical characteristics of enamel in DI, found reduced Vickers hardness of enamel by up to seven times and Young's modulus of elasticity was six times smaller than that of healthy enamel, and concluded that the reduction in these properties could contribute to attrition which can be responsible for the rapid wear of enamel [23].

Historically, treatment of DI patients included extensive restorative therapies ranging from removable prosthetics, to tooth or implant borne full coverage restorations, because once the enamel is lost and the abnormal dentine is exposed, there is a sudden and extensive further loss of the defective dentine and no sufficient tooth structure remains to support other types of conservative restorations.

Improving the tooth colour was one of the main reasons why the patient had presented to us, though she did not want to undergo extensive restorative treatment. Upon further examination of the patient, evident signs of vertical enamel fracture lines and loss of enamel due to wear were also observed. Though her young age, in some of the teeth the wear has already resulted in dentine exposure, thus putting her at risk for further loss of tooth structure [24]. Therefore the main objectives of our treatment were improving her aesthetics whilst protecting her dentition from further wear and fracture. That is why, after classifying the patient as an ACE Class III patient, a modified version of the “three step” sandwich approach [14] was chosen as the most conservative yet comprehensive therapy, in terms of aesthetics and protection of further loss of tooth structure, since the improvement in adhesive technology associated with conservative restorations, has the capacity to increase both anterior and posterior tooth mechanical characteristics. [25, 26, 27, 28, 29, 30].

Though using a more conventional prosthodontic treatment consisting of full coverage crowns, may have masked the pigmentation of her teeth in a more significant way, this novel approach was preferred due to its more conservative nature, and because of the patients young age. Another reason why a conventional prosthodontics approach was not chosen, was the inability to perform root canal therapy, which meant post retained cores under conventional crowns could not be used, and given the possibility that the abnormal dentine may not be used for adhesion in cases of extensive loss of dentine. Therefore adhesion of the restorations directly onto the enamel were chosen because between enamel and dentine, enamel is histologically the more unaffected layer [31] and could be used to a greater extent to bond adhesive restorations, whereas full coverage crowns would rely completely on mechanical retention onto the reduced characteristics of the severely affected dentine.

Recent protocols have been described to adhere new restorations onto previous ceramic restorations in cases of chipping or failure of the porcelain [32]. The correct use of these protocols would also allow us not to have to replace previous tooth borne and implant borne crowns to increase the vertical dimension of occlusion and thus kept the extensiveness and cost of the treatment contained.

Once the treatment therapy was chosen, the next step was to select the adequate materials which would sufficiently fulfill the patients aesthetic and mechanical needs. The masking ability of the minimal thickness of the restorations had to be taken into consideration, since the extent of the discolouration was severe. Recently a study has confirmed the significant influence of the core substrate on the final shade post cementation of the veneer [33].

The alternatives for the upper and lower labial veneers were either feldspathic or lithium disilicate veneers but the chosen material was lithium disilicate as the coping material for ceramic layered veneers since they were the only material which could significantly at-
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tempt to cover the discolouration. Furthermore this material was also available in a number of shades which could be tried in the patients mouth to select the ideal shade. To maintain materials with similar wear coefficients, lithium disilicate palatal veneers were chosen.

In the posterior segments where inlay and onlay restorations had been planned to increase the vertical dimension of occlusion, due to the minimal thicknesses required, CAD/CAM composite inlays/onlays were selected as the material of choice. A recent study comparing the mechanical properties of CAD/CAM composite and ceramic onlays identified composite as the material of choice due to its significantly increased fatigue resistance when compared to ceramic onlays [34].

Conclusion

Given the young age of the patient, it is possible that some maintenance and reintervention especially of the posterior inlays/onlays may have to take place due to chipping or wear of the composite at some later stage, though such interventions are easily and cheaply carried out. The use of lithium disilicate veneers was able to sufficiently mask and improve the aesthetics, strengthen and protect the teeth from further tooth loss. If debonding or other types of failure of the veneers take place they may be also easily carried out.

Apart from the reduced cost of treatment, the advantages of this type of treatment as opposed to a conventional prosthodontic alternative of full crowns, are given by the maintenance of as much normal tooth structure as possible, without having to rely on greatly affected dentine to bear the load of full crowns both in the anterior or posterior segments. Therefore this novel and conservative treatment may be used as a valuable alternative for other patients in similar conditions.

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