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CLINICAL REPORT

A multidisciplinary approach to the functional and esthetic rehabilitation of dentinogenesis imperfecta type II: A clinical report

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ABSTRACT

A 20-year-old woman presented with an unesthetic appearance and severe wear of the anterior teeth. Definitive expected treatment was designed by multidisciplinary combination therapy. Meanwhile, gene sequencing was used diagnostically, and digital smile design (DSD) was used to design the esthetics of the anterior teeth. The 3-month follow-up showed an acceptable outcome. The whole-exon sequencing and direct sequencing of polymerized chain reaction products of dentin sialophosphoprotein were performed, but further sequencing was needed in the repeat region of dentin sialophosphoprotein exon 5 (the data were not shown). DSD assisted and improved the esthetic design of the definitive restoration, communication with the patient, and predictability of the treatment. This treatment demonstrates that the diagnosis and differential diagnosis of patients with dentinogenesis imperfecta (DGI)-II are essential and effective for implementing the definitive treatment plan. Gene screening can be used as an auxiliary method of diagnosing DGI-II. In terms of esthetic design, DSD played an important role in ensuring a satisfactory result. Moreover, a multidisciplinary treatment protocol focusing on prosthodontics, which is different from the traditional treatment of DGI-II and consists solely of prosthodontic rehabilitation, proved highly effective in the esthetic restoration of a patient with DGI-II. (J Prosthet Dent 2019;■:■-■)

Dentinogenesis imperfecta (DGI) is a hereditary disorder of dentin formation following an autosomal dominant pattern of transmission. It affects both formation and mineralization of dentin. This disorder affects both primary teeth and permanent teeth and can also be associated with osteogenesis imperfecta (OI). Shields et al[1] divided DGI into 3 types: DGI-I, associated with OI and resulting from mutations in the collagen type I genes[2]; DGI-II, which manifests clinically the same in teeth as DGI-I but in the absence of OI[3]; and DGI-III, a phenotype characterized by large pulp chambers.[4]

DGI-II is a result of mutations in the dentin sialophosphoprotein (DSPP) gene[5,6] and occurs in approximately 1 in 6000 to 8000 people in the United States.[7] Patients with DGI-II often present with tooth mobility, decreased occlusal vertical dimension, and tooth discoloration characterized by an amber or gray/blue/brown hue.[7] In addition, the enamel in patients with DGI-II tends to chip away from the dentin, exposing dentin dysplasia, which can lead to rapid attrition.[6] Radiographically, DGI-II presents with short, thick roots and pulp obliteration.[8] Crowns on these teeth appear bulbous due to apparent cervical constriction. Given the involvement of numerous symptoms, treatments for patients with DGI-II are complex and difficult. Such treatment involves esthetic reconstruction of

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the anterior teeth, structural protection to prevent the development of caries and wear due to abrasion, and maintenance of the occlusal vertical dimension.[9] Furthermore, differential diagnoses, esthetics, and a multidisciplinary approach should be considered during the treatment process.

As DGI-II is a hereditary disease, gene sequencing can be used to confirm diagnosis and investigate the implicated DSPP mutation(s). Blochzupan et al[6] reviewed 39 mutations in the human DSPP gene that may cause dentin defects. Genetic studies have been hampered in the repetitive region of DSPP exon 5 because of the complex mutation in this region,[10] but McKnight et al[11] cloned and sequenced the DSPP repeat domain and reported a relevant mutation.

Digital smile design (DSD) is a digital tool that amplifies the diagnostic view and can improve communication and enhance predictability throughout treatment.[12] DSD is instrumental in the development of a treatment plan that results in a smile that meets the functional, esthetic, and emotional requirements of the patient.[13] The complexity of rehabilitation of patients with DGI-II necessitates a multidisciplinary team of health-care providers, including an orthodontist, a periodontist, and a prosthodontist.[14,15]

This clinical report illustrates a multidisciplinary treatment of DGI-II to restore and protect esthetic and functional features ranging from stable mastication to facial harmony for long-term satisfaction and delineates some therapeutic guidelines for prosthodontic management.

CLINICAL REPORT

A 20-year-old woman was referred to the Prosthodontics Department of the School and Hospital of Stomatology, Wenzhou Medical University, China. The patient's chief complaint was discolored and unsightly maxillary anterior teeth with morphological defects and severe wear of the mandibular anterior teeth. A panoramic radiograph (Fig. 1A) revealed short roots, pulpal obliteration, cervical constriction, and bulbous crowns. A lateral cephalometric radiograph and tracing (Fig. 1B) showed skeletal class I malocclusion. A family history spanning 4 generations on the maternal side supported a provisional diagnosis of DGI-II (Fig. 1C). The patient was in good physical condition with no evidence of OI.

Pretreatment photographs (Fig. 1D) showed amber crowns, morphological defects of the maxillary anterior teeth, and severe attrition of the mandibular anterior teeth. Gingival hyperplasia was apparent in the anterior maxillary area. The photographs showed a class I malocclusion with excessive horizontal and vertical overlap, labioclination of the maxillary right central incisor, and rotation of the mandibular right second premolar. These clinical manifestations, together with the family history, led to a diagnosis of DGI-II.

To further investigate the diagnosis and etiology of this patient, the exons and exon-intron boundaries of the DSPP gene for this patient were sequenced (data not shown). The study protocol was independently reviewed and approved by the Ethics Committee of the School and Hospital of Stomatology, Wenzhou Medical University. Whole-exon sequencing and direct sequencing of polymerized chain reaction products of DSPP including exon 1-5 were performed for IV-1 (patient), III-2 (another patient in the family), and III-4 (a normal member in the family). The repeat region of exon 5 was not investigated because of challenges associated with accurately sequencing repeat regions. No mutations were discovered in the sequences of exons 1 through 5 (excluding the repeat region), which is consistent with the hypothesis that relevant mutations leading to DGI-II reside in the repeat region of exon 5, as reported in previous studies.[11,16,17]

The definitive objective in this treatment was to restore the function and esthetics of the anterior teeth using a multidisciplinary team approach. The teeth were aligned, and the asymmetrical occlusal curve in the vertical direction was adjusted with orthodontic treatment. Periodontal treatment was used to lengthen the crowns of the maxillary and mandibular anterior teeth. Prosthodontic treatment protected the residual dental tissue from wear and restored the esthetics of the anterior teeth. As the posterior teeth had little attrition and in compliance with the patient's wishes, the posterior teeth were just monitored during the treatment process. Ceramic crowns will be needed if the teeth become damaged.

For esthetic smile planning, a software program (Smile Linker; Hengdasheng) was used. Facial and intraoral photographs of the patient were made (Fig. 2). Based on the facial photograph with a wide smile, the outline of the lip was drawn, and 6 ovate maxillary anterior teeth were chosen according to the facial morphology. The height-to-width ratio was measured, and a dental contour drawing was inserted. The incisal edge was designed following the lower lip line of the intraoral photograph. The tooth color was selected based on the patient's complexion. This design indicated that orthodontic treatment and periodontal surgery were needed to achieve an optimum result.

To align the maxillary and mandibular teeth, brackets were conventionally bonded directly on all teeth (Fig. 3A). As the patient had class I malocclusion with excessive horizontal and vertical overlap, orthodontic extraction was not indicated. Retraction of the maxillary right anterior tooth and rotation of the mandibular right second premolar were chosen. After orthodontic treatment, the photographs (Fig. 3B) showed good intercuspation with a class I molar relationship. The maxillary and mandibular teeth were aligned, and symmetric flattening of the occlusal plane was accomplished.

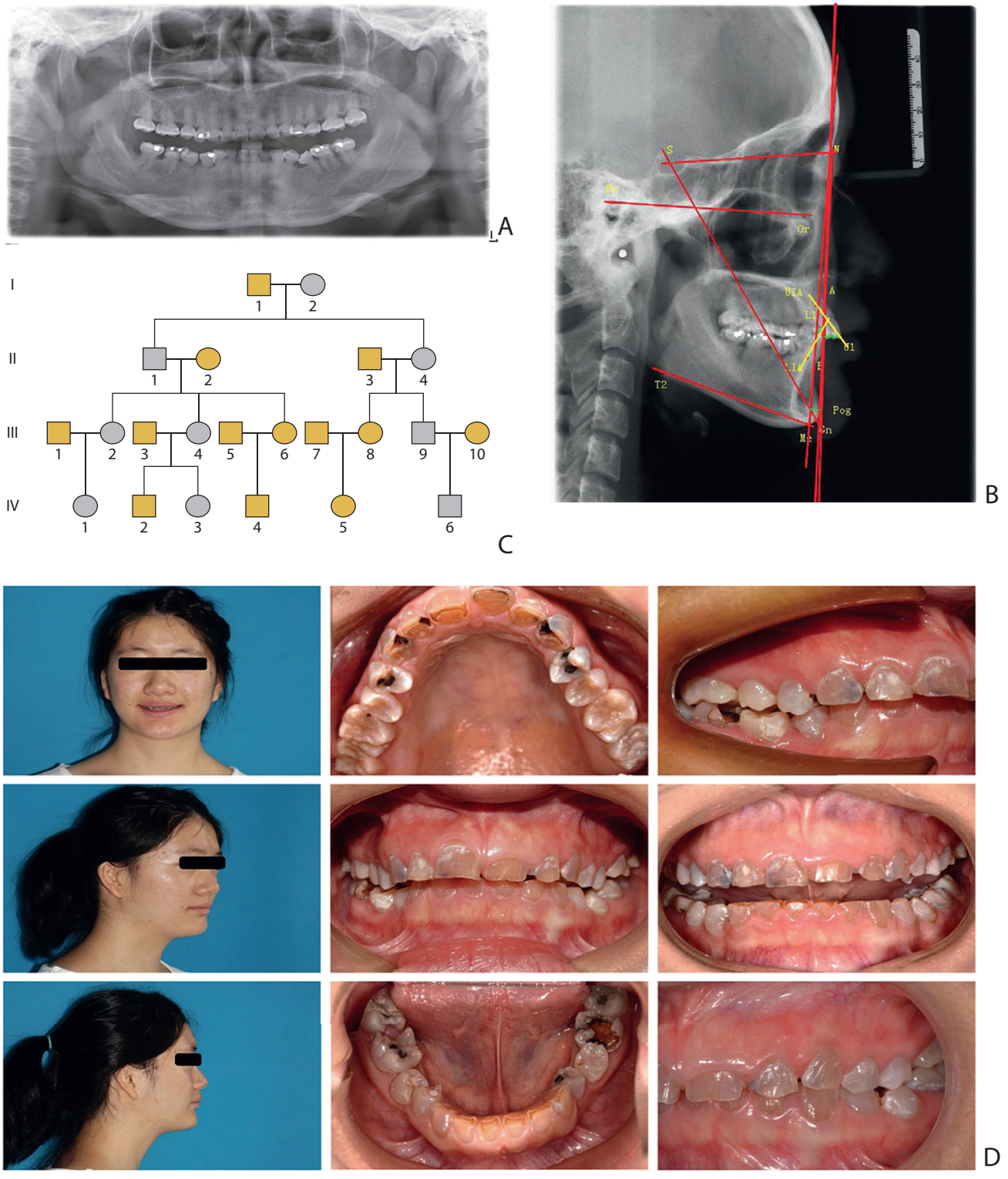

Figure 1. Clinical examination. A, Panoramic radiograph. B, Pretreatment lateral cephalogram and tracing. C, Family history. Each branch of family recorded separately. Patient is IV-1. D, Pretreatment facial and intraoral photographs.

Maxillary and mandibular alginate impressions were made, and gypsum was poured in. The occlusal relationship was recorded and transferred using a facebow (Artex Facebow; Amann Girrbach) and articulator (Artex Articulators; Amann Girrbach). An esthetic diagnostic waxing was performed to evaluate

Figure 2. Process of digital smile design.

Figure 3. Orthodontic treatment. A, Intraoral photographs during orthodontic treatment. B, Intraoral photographs after orthodontic treatment.

the definitive outcome and was an effective visual communication tool for all partners involved in this treatment (Fig. 4).

According to the DSD result and esthetic waxing, crown-lengthening surgery and gingivoplasty were needed in the maxillary anterior teeth owing to the gingival hyperplasia and asymmetry of the gingival margin to ensure enough crown-height retention. Crown-lengthening surgery was also indicated for the worn mandibular anterior teeth to ensure adequate crown retention. Crown-lengthening surgery, gingival recontouring, and osteotomy were performed on the

Figure 4. Waxed diagnostic cast.

Figure 5. Periodontal surgery.

maxillary anterior teeth (Fig. 5). As the attached gingiva was limited in the mandibular anterior region, an apically repositioned flap surgery was performed with osteotomy (Fig. 5). After periodontal treatment, the gingival contours of the maxillary and mandibular incisors were more esthetic, gingival exposure was reduced when smiling, and the crowns were lengthened.

After a month of periodontal treatment, trial restorations were made using silicone rubber (Silagum-Putty; DMG Chemisch-Pharmazeutische Fabrik GmbH) and bis-acrylic resin (3M ESPE Protemp II; 3M Deutschland GmbH) (Fig. 6A). Predetermined depths were prepared using diamond rotary instruments according to the thickness required for each of the ceramic crowns (Fig. 6B). The esthetics of the definitive restoration were evaluated and adjusted for optimal occlusion (Fig. 6C). Ceramic definitive restorations were provided to protect the residual dental tissue from wear. Posttreatment photographs showed that the definitive restorations were satisfactory, and the patient was satisfied with the treatment result (Fig. 6D). No postoperative complications were detected after 3 months, and the patient was satisfied with the esthetic and functional outcomes (Fig. 7). Figure 8 illustrates the entire treatment process.

DISCUSSION

This treatment emphasizes issues surrounding DGI-II. Diagnosis and differential diagnosis must differentiate among the 3 distinct types of DGI. DGI-I and DGI-II are similar in clinical appearance, radiographs, and histology, but patients with DGI-I also suffer from OI, which requires a more comprehensive treatment management.[3] This patient was diagnosed with DGI-II as no evidence of OI or other types of DGI was found.

Evaluation of esthetics is subjective to and influenced by factors including characteristics of culture, gender, body type, and age. Therefore, the esthetic design of the smile and a personalized treatment plan are important.[14] Smile Linker, a convenient and precise DSD software program for iPad, was used to design the treatment plan. In showing the design results, treatment strategies, prognosis, and recommendations, this software allowed objective communication with the patient, facilitated acceptance or modification of the design, and assisted the clinician throughout the treatment process.[12]

After the DSD design, a multidisciplinary approach was key in achieving favorable clinical outcome. The use of orthodontic treatment for alignment of teeth is controversial for patients with DGI because of the risk of

Figure 6. Prosthodontic treatment. A, Evaluation of suggested anatomy and esthetics for definitive restorations. B, Tooth preparations under trial restorations.

Figure 6. (*continued*). C, Interim restorations. D, Facial and intraoral views after prosthodontic rehabilitation with lithium disilicate ceramic crowns.

Figure 7. Follow-up after 3 months.

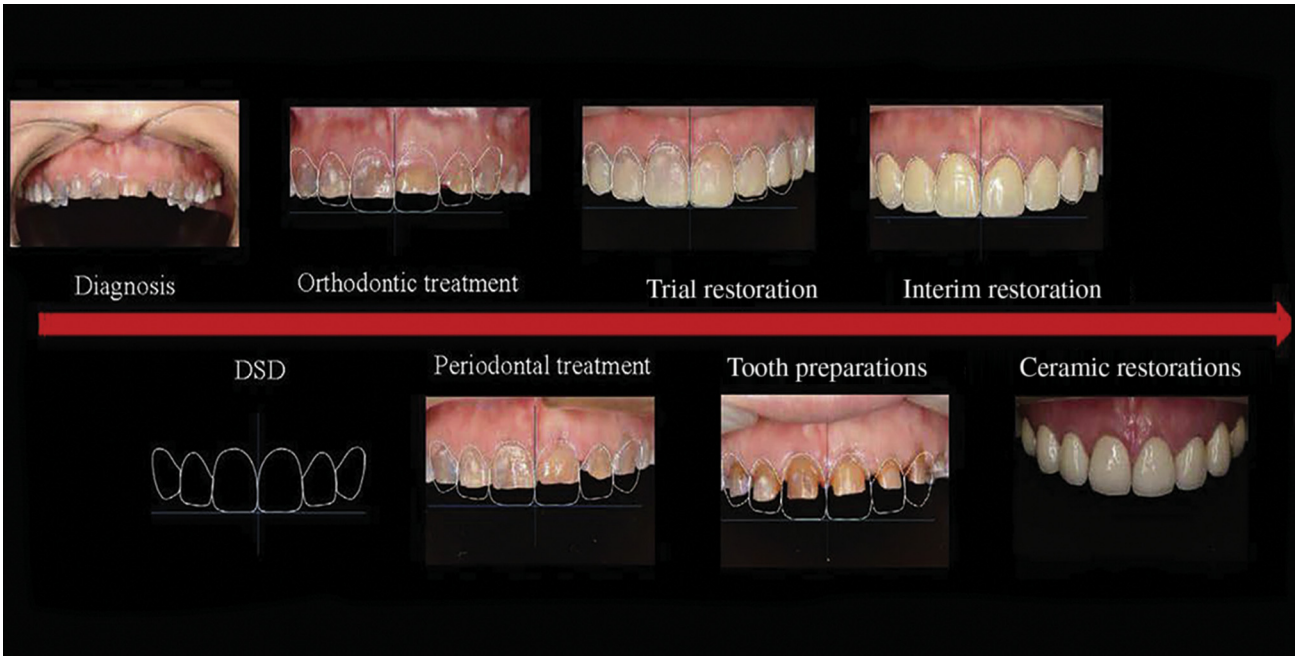

Figure 8. Entire treatment process. DSD, digital smile design.

enamel fracture. However, Roh et al[15] reported excellent results when treating a patient with DGI-II with asymmetric extractions and treatment mechanics in orthodontics. Because of the malalignment of teeth, orthodontic treatment was needed for the present patient. However, the horizontal and vertical overlap could not be altered because the retraction space could not be retained and the situation did not permit

orthodontic tooth extraction. Moreover, root apices may perforate the bony cortex if the horizontal and vertical overlap of anterior teeth is modified to provide normal occlusion. Given the potential for bonding failure or enamel loss after rebonding or debonding because of enamel fractures associated with DGI-II, resin cement was used on all teeth.[18] During orthodontic treatment, brackets bonded successfully to the tooth surface, and enamel fracture was not observed, consistent with the findings of Roh et al.[15]

As this patient presented with gingival hyperplasia and asymmetry of the gingival margin in the maxillary anterior region, crown-lengthening surgery and gingivoplasty were needed to meet the esthetic treatment goals and allow for sufficient crown retention. Providing adequate restoration space was difficult because of the worn mandibular incisors. However, the occlusal surfaces of the molar and premolar teeth were unworn, and the temporomandibular joint was normal. Therefore, increasing the occlusal vertical dimension was unsuitable for this patient. Gingivectomy to increase the length of teeth was also inappropriate because of the limited attached gingiva. As a result, apically repositioned flap surgery and osteotomy were chosen for crown lengthening.

During the prosthodontic rehabilitation, the dental laboratory transferred the virtual restoration design and patient preferences to dental casts for diagnostic waxing.[19] Interim restorations were fabricated based on the diagnostic waxing to further improve visualization of the digital restoration design.[12] Enamel was preserved by guided tooth preparation. Lithium disilicate ceramic crowns were chosen to protect the residual dental tissue from wear and to restore the esthetics of the anterior teeth.[20]

SUMMARY

The diagnosis and differential diagnosis of patients with DGI-II are essential and effective in implementing a definitive treatment plan. In esthetic design aspects, DSD can play an important role in achieving a satisfactory result. Moreover, a multidisciplinary treatment protocol is highly effective in esthetic restoration for a patient with DGI-II.

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