PROSTHETIC TREATMENT IN A PATIENT WITH OSTEOGENESIS IMPERFECTA

By

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ABSTRACT

Osteogenesis imperfecta is a syndrome of interest to dentists because dentinogenesis imperfecta is frequently associated with not only bone disease but also tooth disease. Specifically, these patients usually have many dental problems such as 1) high incidence of tooth fracture, enamel chipping and cracking, 2) high caries activity, 3) inability or difficulty of endodontic treatment, 4) masticatory disturbance and 5) esthetic dissatisfaction.

This paper presents a case report of a patient with osteogenesis imperfecta and dentinogenesis imperfecta who was provided prosthetic treatment, including endodontic treatment of the obliterated teeth.

INTRODUCTION

Osteogenesis imperfecta is a serious hereditary disease caused by mesodermal disorders. The chief clinical characteristics of this disease are fragility of the bone, blue coloration of the scleras, and deafness. In the stomatognathic system, this disease is closely related to dentinogenesis imperfecta and the oral condition is clinically characterized by chipping and cracking of enamel with a rapid wearing down of teeth and opalescent color, giving rise to early loss of teeth and subsequently to diminished vertical dimension of occlusion.

Therefore, most of these patients usually complain of masticatory disturbance and esthetic dissatisfaction because of the foregoing dental problems. However, it is difficult for these patients to retain the prosthesis on the teeth because of the short length of clinical crown, soft dentine, and inability or difficulty of endodontic treatment due to the obliterated pulp chamber and root canal.

This paper describes the prosthetic treatment provided to a patient with osteogenesis imperfecta and dentinogenesis imperfecta, including endodontic treatment for the obliterated pulp chamber and root canal, to bring about improvement of masticatory function. In addition, an evaluation was made of masticatory function electromyographically during the course of treatment.

CASE REPORT

General Condition

The patient, a 20-year-old female, was referred to our dental clinic in October, 1978 for improvement of masticatory disturbance and esthetic dissatisfaction. Review of the patient's medical history disclosed osteogenesis imperfecta...
congentia that began from birth with multiple fractures of the leg. In the patient's pedigree, there were no cases of osteogenesis imperfecta in three generations (Fig. 1).

Physical examination revealed several typical features of this disease including diminished stature with scoliosis, bowing of the legs, and long slender arms. The neck showed distinct webbing and was very short. The ligament laxity was normal. The scleras were blue, but deafness was not found.

Oral Condition

The mandibular and maxillary anterior teeth exhibited dentinogenesis imperfecta with characteristic opalescent dentine reddish-brown in color. In the maxillary anterior teeth 212 were fractured with only the root remaining and 11 was missing. The mandibular and maxillary posterior teeth also exhibited dentinogenesis imperfecta, and 616, 76167 were missing. The gingival tissue was slightly red and swollen, and the oral hygiene in this patient was very poor. In addition, the tongue was disproportionately large (Fig. 2).

Roentgenographic examinations of the teeth revealed the outline of the crowns to be bell shaped with slender roots of normal length. The root canals were slightly visible, but the pulp chambers and root canals were obliterated and filled. The periodontium appeared normal, but the mandibular base bone was thin and porous (Fig. 3).

Parietal bone expansion gave a tapered appearance to the head and face. Mandibular prognathism and maxillary retraction in profile were observed, which was verified by cephalometric analysis (Fig. 4).

Fig. 1. Chart of the pedigree of the patient in three generations. ♀ : patient, ♂ : deceased. Figures denote age in years. Note absence of affected persons in the family.

Fig. 2. Frontal oral view of the patient before treatment. Note dentinogenesis imperfecta in the anterior teeth.

Fig. 3. Roentgenograph of the patient. (A) : dental, (B) : pantomograph. Note obliteration of the pulp chambers and root canals, bulbous shape of the crown, constriction of the cervical portion of these teeth, and thinness and porosity of the mandibular base bone.

Fig. 4. Cephalometric roentgenograph (A) and its analysis (B) showing soft tissue and bony landmarks indicating mandibular prognathism and maxillary retraction.
Prosthetic Treatment in Osteogenesis Imperfecta

Treatment Procedure
It was considered that the patient's profile (mandibular prognathism and maxillary retrusion) could be best improved by combination of joint surgery (bilateral osteotomy of the ramus) and prosthetic treatment. As this patient did not consent to surgical treatment, prosthetic treatment was given to correct only the masticatory disturbance.

Prosthetic treatment in this patient required such special considerations as 1) possibility of teeth fractures, enamel chippings and crackings, 2) high caries activity, 3) inability or difficulty of endodontic treatment, 4) decreased vertical dimension, 5) decreased contact areas in centric occlusion and 6) mandibular prognathism and maxillary retrusion. In view of these, full mouth reconstruction with the crown and bridge was considered to be more appropriate for this patient than with overdenture. It was expected that 3) could be solved by vital amputation at the low level of the pulp.

The following procedures were employed in the prosthetic treatment: 1) vital amputation at 2112, 2) extension of the vertical dimension (about 5 mm) with temporary bilateral posterior metal bridges to maintain the new vertical dimension for a month as evaluated by mandibular kinesiograph, 3) construction of final bilateral posterior bridges and anterior porcelain fused to metal bridge and crowns. The frontal oral view after the prosthetic treatment is shown in Fig. 5.

Following each construction by the usual procedure, masticatory performance and EMG activities in this patient were evaluated to estimate the level of masticatory function.

Methods and Results of Masticatory Performance and EMG Activities
Masticatory performance was measured by using the method described by Manly and Braley. Nine grams of peanuts were divided into three portions. The patient was instructed to chew each portion for twenty masticatory strokes with the habitual chewing side and rate and to eject the chewed peanuts into a 10 mesh screen. This was repeated for five times. Peanuts remaining on a 10 mesh screen and those passing through a 10 mesh screen were dried in an oven at 100°C for three hours and then in a desiccator for two hours. They were weighed to calculate the masticatory performance.

EMG activities were recorded from both the masseter and anterior temporal muscles in a conventional manner with surface electrodes (Beckman Instruments, Inc.) connected to a biophysical amplifier (Nihon Koden Kogyo Co., Ltd.) having an input impedance of 5 MΩ. Direct EMG activities were recorded by a photocorder (Yokogawa Electric Work. Ltd.). The patient was instructed to chew two grams of peanuts with the habitual chewing side and rate. The duration, interval and cycle of the EMG activities were measured and calculated during this chewing effort. These procedures were repeated for five times.

The masticatory performance of this patient was extremely poor before the prosthetic treatment, but remarkable improvement was observed following prosthetic treatment. The values of masticatory performance were 4.0 ± 1.4% before treatment, 14.5 ± 0.4% during temporary bridge, and 22.7 ± 2.0% following prosthetic treatment, respectively.

EMG activities from the masseter and anterior temporal muscles before the prosthetic treatment were extremely unstable and not rhythmical. Especially, the amplitude of the EMG activities from the masseter muscle was so small that it could not be detected without difficulty and therefore the EMG activities from the anterior temporal muscle were mainly selected for observation. After completion of the prosthetic treatment, the EMG activities from both the masseter and anterior temporal muscles became remarkably stable and rhythmical and the chewing strokes decreased to two-

Fig. 5. Frontal oral view of the patient after the treatment.
according, the former being transmitted as autosomal dominant trait and the latter as autosomal recessive. The clinical diagnosis of this case was osteogenesis imperfecta congenita because of the onset pattern of this disease and the absent of any expression of its phenotype in the patient's pedigree.

Since osteogenesis imperfecta is basically a disturbance of the mesodermal tissue, dentinogenesis imperfecta is a common companion of this disease in the stomatognathic system. Dentinogenesis imperfecta is characterized by a peculiar tooth color, severe attrition, and obliterated pulp. It can occur in both the deciduous and permanent teeth. The teeth are opalescent or violet and the enamel tends to chip or shear away from the dentine. Histologically, chemically and physically, the enamel is normal, but the dentine is poorly calcified. Dentinal fibers are few in number but of a large diameter and are arranged haphazardly and occasionally appear at right angles to the normal direction. In our patient the teeth exhibited dentinogenesis imperfecta and the soft tissues, particularly gingiva, were normal except for slight reddening and swelling due to poor oral hygiene. It has not been reported that soft tissues are injured by this disease.

Treatment of these patients is difficult. Helmers and Finn has given four objectives in treatment of patients with dentinogenesis imperfecta: 1) to maintain vertical dimension; 2) to maintain arch length; 3) to ensure the integrity of erupting teeth and 4) to make necessary esthetic correction. To achieve these objectives most authors favor early treatment with metal crown and acrylic crown for these teeth. When attrition is extensive with many missing teeth, they favor placement of an overdenture. The main problem with our patient was masticatory disturbance and esthetic dissatisfaction. For correction of esthetic dissatisfaction overdenture might be one of the possible solutions, but for this patient overdenture was not acceptable because of severe disharmony of the profile and necessity of large retentive force. Therefore, from a functional viewpoint of mastication we decided that correction was made by crown and bridge with the aim of preventing fracture, enamel chipping and cracking, caries, and other dental problems.

DISCUSSION

According to the literature, osteogenesis imperfecta is classified into osteogenesis imperfecta tarda and osteogenesis imperfecta congenita, the former being transmitted as autosomal dominant trait and the latter as autosomal recessive. The clinical diagnosis of this case was osteogenesis imperfecta congenita because of the onset pattern of this disease and the absent of any expression of its phenotype in the patient's pedigree.

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in the remaining teeth in the future.

In the correction procedure with crown and bridge of this patient, it was not possible to construct the crown and bridge at the proper place of the anterior teeth because the remaining root was at 212 and could not accept placement of long pins to enhance the retentive force. Moreover, the pulp chamber and root canal of the teeth might have become obliterated by calcification, and although exposed the pulp did not cause reduced vitality of the teeth. Therefore, it was necessary to make a post-core to retain the crown and bridge. However, since exact pulpectomy and root canal treatment could not be made, vital amputation at the low level of the root canal was undertaken to resolve the difficulty or inability of endodontic treatment. No prognostic problem developed after vital amputation. Therefore, it may be suggested that vital amputation at the low level of the root canal is a valuable therapy in making use of the obliterated teeth as abutment teeth, although further histological and pathological investigations should be made for confirmation.

During the prosthetic treatment, 417 were fractured and there was a possibility that the remaining teeth might be fractured even by slight force. Therefore, all the remaining teeth should be covered with the crown and bridge to avoid fracture, caries, and tooth loss in the future. It is also important to take care of occlusion in placing the crown and bridge.

After the prosthetic treatment the masticatory function was remarkably improved with masticatory performance increasing about 6 times that prior to treatment. The EMG activities after the treatment were more stable and rhythmic than those before the treatment, and the amplitude of the EMG activities after the treatment increased to about 150% that before the treatment. In fact, the patient claimed that she could chew anything better after the treatment. These results suggest that the patient was able to wear and accustom herself satisfactorily to the prosthesis. The improvement of masticatory function was mainly due to increased contact area in the centric occlusion, rehabilitation of the occlusal arch and vertical dimension, and activation of masticatory function.

Finally, the prognosis in osteogenesis imperfecta can not be predicted in this case because of the inherent weakness of the teeth, high caries activity, and uncertain periodontal conditions and therefore special care and further detailed investigation are indicated in this patient.

REFERENCES