Dentinogenesis imperfecta (DI) type 2 is a disease inherited in a simple autosomal dominant mode. As soon as the teeth erupt the parents may notice the problem and look for a pediatric dentist’s advice and treatment. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth and occlusion and improve esthetics.

The purpose of this article is to present the objectives, treatment options, and problems encountered in the treatment of DI in the early primary dentition. A two-stage treatment of a toddler under general anesthesia is described and discussed.

This paper recommends for severe cases of DI two treatment stages performed under general anesthesia. Stage 1 is early (around age 18-20 months) and is directed to covering the incisors with composite restorations and the first primary molars with preformed crowns. Stage 2 (around age 28-30 months) seeks to protect the second primary molars with preformed crowns and cover the canines with composite restorations. (Pediatr Dent 23:232-237, 2001)

Dentinogenesis imperfecta (DI) or hereditary opalescent dentin, was first described in the late 19th century. It is a localized mesodermal dysplasia affecting both the primary and permanent dentition. The disease is inherited in a simple autosomal dominant mode with high penetrance and a low mutation rate. The reported incidence in the USA is 1:8000 births. Shields et al proposed three types of dentinogenesis imperfecta: DI type 1 is associated with osteogenesis imperfecta. DI type 2 has essentially the same clinical radiographic and histological features as DI type 1 but without osteogenesis imperfecta; DI type 3 is rare and is only found in the triracial Brandywine population of Maryland. It has been suggested that DI type 2 and DI type 3 are different expressions of the same gene.

Clinically, with DI both dentitions are affected. The color of the teeth varies from brown to blue, sometimes described as amber or gray, with an opalescent sheen. The enamel may show hypoplastic or hypocalcified defects in about one-third of the patients and, in an affected patient, tends to crack away from the defective dentin. The exposed dentin may undergo severe and rapid attrition.

Radiographically, the teeth have bulbous crowns with constricted short roots. Initially, pulp chambers may be abnormally wide and resemble “shell teeth,” but they will progressively obliterate. Histologically, the enamel, although normal in structure, tends to crack. The dentin-enamel junction is not scalloped. In most cases the structure of the mantle dentin is normal, whereas the dentinal tubules of the circumferential dentin are coarse and branched and the total number of tubules is reduced. The presence of an atubular area in the dentin with reduced mineralization and a reduced number of odontoblasts are consistent findings. Pulpal inclusions and much interglobular dentin are also frequent.

The biochemical characteristics of the dentin include a collagen defect and a primary defect in the calcifying matrix. Takagi and Sasaki suggested that the dentin in DI type 2 is deficient in the phosphorus ion, which is important in the early stage of odontoblastic differentiation and its mineralization. Susuki et al published a case describing DI type 2 with absent enamel prisms and abnormal mantle dentin.

The purpose of this article is to present the objectives, treatment options and problems encountered in the treatment of DI in the early primary dentition. A two-stage treatment of a toddler under general anesthesia is described and discussed.

Dental treatment

In DI, the primary dentition appears more severely affected than the permanent dentition, evidenced by rapid wear of the teeth. This attrition may cause pulpal involvement with dental abscesses, and the short, constricted roots might break under load, thus necessitating extraction. The severe attrition may result in a rapid decrease in the occlusal height. In the early primary dentition, these appear to be the most immediate problems, and soon after eruption it is generally necessary to protect the primary molars with stainless steel crowns.

In the restorative treatment of pediatric patients, glass ionomer with fluoride-releasing and chemically attaching materials are recommended for occlusally non-stressed areas. An acid etch technique followed by composite restoration is proposed as an alternative for restoration of the anterior teeth. Polycarbonate crowns may offer an alternative for the restoration of the anterior primary teeth. An acrylic overlay denture, resting over the remnants of crowns and roots of the primary dentition, also has been used successfully.

Wright has stated that the dental approach for managing dentinogenesis imperfecta will vary with the severity of the clinical expression, while intracoronal restoration and bonded veneers for anterior teeth may be acceptable in mild cases, they might not last in severe cases exhibiting enamel fracturing and rapid wear.
The objectives of early treatment of DI in the primary dentition are as follows:

1. Maintain dental health and preserve vitality, form, and size of the dentition.
2. Provide the patient with an esthetic appearance at an early age, in order to prevent psychological problems.
3. Provide the patient with a functional dentition.
4. Prevent loss of vertical dimension.
5. Maintain arch length.
6. Avoid interfering with the eruption of the remaining permanent teeth.
7. Allow normal growth of the facial bones and temporomandibular joint (TMJ).
8. Establish a rapport with the patient and the patient’s family early in the treatment.

Treatment of the mixed and permanent dentition is challenging and frequently demands a multidisciplinary approach. Collaboration of the pediatric dentist with a prosthodontist and an orthodontist is often imperative. Although caries is not a major concern in most cases, strict oral hygiene instructions and preventive treatment is important in order to prevent caries from adding to existing problems. Frequently, there is a need to reestablish the vertical dimension of occlusion in order to restore the occlusion in the mixed and permanent dentition. Prosthetic restoration combined with orthodontic treatment may be advantageous, and evaluation of the occlusion prior to initiation of treatment is advised. 18
Many treatment modalities have been suggested: overdentures, stainless steel crowns, jacket crowns, pin-retained cast gold “thimbles” under acrylic resin crowns, stainless steel crowns with acrylic facing, and simple, removable appliances.\textsuperscript{16,19-22} A combination of partial dentures and prosthetic crowns on the anterior teeth has also been described.\textsuperscript{23} Indirect resin crown (IRC) technique to restore the maxillary incisors has been used to treat a patient with severe enamel hypoplasia and attrition. This technique may offer esthetic, affordable, and long-lasting intermediate restoration.\textsuperscript{24} Orthodontic treatment has been successfully performed in patients with different degrees of DI.\textsuperscript{25-27} In less severe cases of DI, carbamide peroxide bleaching has been successfully used to treat discoloration.\textsuperscript{28}

**Case report**

ML, an 8-month-old Caucasian girl, was brought by her parents to Department of Pediatric Dentistry at the Hadassah School of Dental Medicine in Jerusalem, Israel. The parents had noticed that the newly erupted lower incisors were grayish. The child was healthy, but the family history revealed that the father, his cousin, and their parents had DI. The father was treated in the Hadassah clinic 32 years ago when he was 7 years old. His cousin was 14 years old when treated with removable overlay bridges. The oral examination of the child revealed partially erupted incisors with a brown blue color. The teeth were very small. The radiographic examination revealed poorly calcified incisors, exceptionally thin dentin, with wide pulp chambers resembling “shell teeth” (Fig 1).

At the follow-up examination at age 14 months, severe attrition of the erupted teeth was noticed. That necessitated early intervention to intercept the developing malocclusion and tooth attrition and in order to preserve the vitality and form of the teeth. The anxious parents were interested in beginning treatment as early as possible. The parents expressed concern that the child and her peers would notice the discolored teeth. Since the father was edentulous and had undergone much suffering, he was very concerned that his daughter would not encounter the same problems. It was decided to cover the teeth with crowns in two stages:

**Stage 1** – as soon as possible, (at age 18-20 months), to cover the incisors with composite restorations and the first primary molars with preformed crowns.

**Stage 2** – (at age 28-30 months) to protect the second primary molars with preformed crowns and to cover the canines with composite restorations.

At 20 months of age the child was admitted to the operating room and treatment was performed under general anesthesia. Close observation of the teeth revealed severe attrition of the primary first molars. Enamel existed only in the unerupted subgingival part of the crowns of the primary first molars. The incisors were spaced with extremely small crowns and with an exceptionally thin buco-lingual dimension (Figs 2-3). Radiographically, the pulp chambers were very wide, with thin dentin, resembling “shell teeth” (Fig 4).

An attempt was made to fit polycarbonate crowns and preformed polymer glass crowns (Glasstech, Austin, Tx.) for the anterior teeth, but both were too large labiolingually. In order not to weaken the teeth and to prevent pulp exposure, no reduction of tooth substance was performed. After total etching of the incisors with 37% phosphoric acid for 30 seconds, the teeth were coated with two layers of Single-Bond (3M). Pedoform strip crowns (3M) were filled with composite Z-100 (3M) and cured for 80 seconds.

The preparation of the primary molars presented us with other problems: Short tooth crowns due to insufficient eruption and attrition, and thin, short, fragile roots. The cervical enamel was reduced slightly using a tapered diamond, and care was taken not to reduce the rest of the crown. Unitek stainless steel crowns (3M) were shortened and individually adapted. The crowns were glued with Glass Ionomer Cement, using only moderate pressure (Fig 5). During the follow-up over the next 6 months, some of the composite restorations on the anterior teeth chipped away and needed replacement. The child was re-admitted and the restorations were replaced under sedation.

At the age of 30 months, the second stage of treatment was conducted under general anesthesia. Preoperative clinical view (Fig 6), shows some of the composite restorations that were missing or chipped away and the extremely small clinical crowns of the primary canines that underwent rapid attrition.
Preoperative radiographs done at that time (Fig 7) demonstrated the pulp obliteration of the primary incisors and the attrition of the canines. Only minimal reduction of tooth substance was performed on the treated teeth. The second primary molars were covered with stainless steel crowns, and the canines were covered with pedoform strip crowns (Fig 8). The six-month followup examination revealed the restorations to be esthetically acceptable and functional.

Discussion

The importance of restoring dental defects associated with DI is obvious. There is no clear answer as to the best time for initiating treatment. The literature abounds with case reports and treatment modalities for DI; in general, the recommendation is to start treatment as early as possible.\textsuperscript{13,17} The rational use of general anesthesia or pharmacotherapeutic regimens at an early age enables us to safely utilize state of the art restorative techniques in the treatment of young patients suffering from hereditary dental anomalies.\textsuperscript{29} However, there are only two case reports describing treatments of the very young child.\textsuperscript{30,31} Daugard-Jensen and Nielsen described 3 cases, the youngest being a 19-month-old with DI type 1.

The initial treatment was with stainless steel crowns on the molars which were replaced later with cast gold crowns due to severe attrition. The cast gold crowns were not retentive and parapulpar pins were prepared to improve retention. The anterior teeth were restored in one case with polycarbonate crowns and with stainless steel crowns in another case.\textsuperscript{30} Torija and Quijada described the dental treatment of a 3-year-old child with DI type 1. The treatment included covering the posterior teeth with stainless steel crowns and the anterior teeth with stainless steel crowns with open face composite restorations.\textsuperscript{31}

Treating a child at such an early age raises certain questions and problems. The main dilemmas involved in the early treatment of DI include:

1. High cost.
2. Risk of general anesthesia.
3. Postponing treatment could result in better cooperation and obviate the need for a general anesthetic.
4. There are technical problems in restoring teeth with minimal clinical crowns and wide pulp chambers.

However, the advantages of early treatment outweigh the disadvantages:

a. Early treatment can prevent/intercept possible problems: loss of the vertical dimension, possible TMJ problems, and arch length complications, thus providing the patient with a healthy and functional dentition.

b. The importance of an esthetic look cannot be ignored. The smile is very important for socialization and for the child’s “self concept” from an early age and for establishing a positive “basic reality.”\textsuperscript{32-34}

c. The concern of the parents and their wishes should not be overlooked.

The problem of wide pulp chambers and thin dentin in this very young patient presented great difficulty in the preparation of the teeth. In order to receive maximal retention for the bonded composite, some preparation of the teeth eventually was necessary. However, in the first stage of treatment this was not done. It is possible that this resulted in a low retention rate of some of the composite restorations. However, the incisors were still protected from pulp exposure and further attrition until the next stage of treatment, when a few of the anterior restorations were redone.

The clinical severity of DI needs to be assessed when developing a treatment plan. When the disease is more severe, the biochemical properties of the enamel and dentin are compromised,\textsuperscript{11} and this can affect adhesion strength.

In spite of the success described earlier with composite restorations in treatment of DI in permanent teeth,\textsuperscript{14} success might not be the same in severe cases of DI in the primary
dentin. In such cases, it may be better to prepare the teeth with a diamond bur, take off all of the enamel that might crack off in the future, and then bond the composite restoration to the dentin. Rada and Hasiakos have suggested using glass ionomer cements and dentinal adhesives as the first layer, with a composite restoration on top, in the treatment of Amelogenesis Imperfecta. They hypothesized that chemical binding might improve the durability of the restoration.

In less severe cases, where normal enamel exists, mechanical preparation of the enamel may be beneficial in reducing the apismatic layer which exists even in normal incisors. This may expose the prismatic enamel and improve mechanical retention.

In the future, and with better patient cooperation, it may be possible to suggest transitional treatment with IRC. This intermediate modality of treatment, combined with chemical adhesion of composite resin cement and the mechanical adaptation of a well-adjusted crown may solve retention problems.

Conclusions
1. Early diagnosis and treatment of DI is recommended, as it may prevent or intercept deterioration of the teeth and occlusion and improve esthetics.
2. In severe cases of DI, two treatment stages of the primary dentition under general anesthesia are recommended.
3. In moderate cases, a one-stage treatment of the primary dentition at age 30 months may be justified.
4. Composite restoration in severe cases of DI might not prove durable.
5. Long term followup is imperative in order to intercept complications and adjust the treatment to the changes of the dentition and occlusion.

References

ABSTRACTS OF THE SCIENTIFIC LITERATURE

Effectiveness of polymerization in composite restorations claiming bulk placement

The manufacturers of two composite resins (Ariston pHc and Surefill) claim that these composites can be effectively cured to depths of 4-5 mm in 40 seconds of exposure to visible light, and suggest the possibility of bulk placement of the material in cavity preparations. The impact of increasing cavity depths and light-source exposure times upon the effectiveness of polymerization of composite resins can be evaluated by hardness testing. The objective of this in vitro study was to evaluate the hardness gradient between top and bottom surfaces of composite specimens of various depths after different light-exposure times. The data presented demonstrate that increasing cavity depth and increment size result in a significant decrease in the effectiveness of polymerization for all exposure times.

Comments: This study demonstrates that increments of the two composites evaluated should be no greater than 2 mm to obtain uniform and maximum cure. Therefore, incremental technique should be used whenever a cavity preparation is larger than 2 mm. JN

Address correspondence to: Dr. A.U.J. Yap, Dept. of Restorative Dentistry, Faculty of Dentistry, National University of Singapore, 5 Lower Kent Ridge Road, Singapore 119074, Republic of Singapore.

21 references

Bitewing radiography and fiber-optic transillumination in caries diagnosis

This study evaluated the performance of two techniques for the diagnosis of approximal caries in low caries prevalence populations. Only clinical studies that evaluated the two techniques were selected from the literature for analysis. 2 x 2 contingency tables were determined from the selected studies from which one overall contingency table was calculated. The cut-off for decay was dentinal caries. Three situations were examined: Bitewing radiography specificity is 1, Transillumination specificity is 1 and both specificities are equal. Under these conditions, bitewing sensitivity ranged from 1.00-0.71±0.1. Fiber optic transillumination sensitivity ranged from 0.7±0.1 and 0.50±0.02 and the specificity of both techniques was between 1.00 and 0.99. The authors conclude that as in use today, fiber optic transillumination is inferior to bitewing radiography.

Comments: Taken in total, the literature does not support this alternative method as the sole technique for the detection of approximal decay. CH

Address correspondence to: J. Vaarkamp, St. Jude Children’s Research Hospital/Radiation Oncology, 332 North Lauderdale St., Memphis TN 38105-2794.

22 references