Case Report

# Dental Management of a Child with Dentinogenesis Imperfecta: A Case Report

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## Abstract

Dentinogenesis imperfecta (DI) is a hereditary dentin defect caused by an autosomal dominant mutation in dentin sialophosphoprotein gene. Defective dentin development results in discolored teeth that are prone to wear and fracture. Early diagnosis and proper treatment are necessary to achieve better functional and esthetic results and minimize nutritional deficiencies and psychosocial distress. In order to prevent excessive loss of tooth structure, placement of stainless steel crowns (SSCs) on deciduous and young permanent posterior teeth is recommended as soon as such teeth erupt. This clinical report presents the clinical manifestations and management of a 3.5-year-old child diagnosed with DI type II.

**Keywords:** Dentin; Dentinogenesis Imperfecta; Tooth, Deciduous

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## INTRODUCTION

Dentinogensis imperfecta (DI) is the most common hereditary disorder of dentin formation (1:8000) [1], which is inherited as an autosomal dominant trait [2] with high penetrance and a low mutation rate [3]. In 1908, DI was diagnosed for the first time as a defect, which is predominantly due to abnormal dentin [4]. Mutations in dentin sialophosphoprotein gene, located at locus 4q12-q21, [5] cause defects in dentin sialo-phosphoprotein and dentin phosphoprotein, which are responsible for 50% of non-collagenous structure of dentin [6].

Dentinogensis imperfecta has been classified by Shields et al, [7] into three categories: Type I is associated with osteogenesis imperfecta. Primary teeth are more severely affected than permanent teeth. In Type II, which is also known as heredity opalescent dentin, primary and permanent dentition are equally affected. Type III is rare and most commonly affects the permanent dentition, and gives them a shell-like appearance. It has only been reported in patients from Brandywine, where there is a large population of patients suffering from this condition [7,8].

The most common clinical manifestations of DI are tooth discoloration (varying from amber-like translucent gray or brownish purple to yellowish brown) [9,10] and excessive tooth wear. The enamel might peel off to leave dentin exposed, making it susceptible to severe and rapid decay [10]. Radiographically, teeth have normal enamel radiodensity and thickness [8]. Other features of DI include bulbous crowns with marked cervical constrictions and partial or total precocious obliteration of pulpal space [11]. Significant decay can be seen over a short period of time [12]. Histologically, dentinal tubules are sparse, irregular, larger in diameter, and often have large areas of uncalcified matrix [3].

Early diagnosis and proper treatment are necessary to achieve better functional and esthetic results. Most patients with DI require a comprehensive multidisciplinary treatment plan, which depends on the patient's age at the time of diagnosis, clinical condition, severity of disease, expectations, compliance and available resources [1,13].

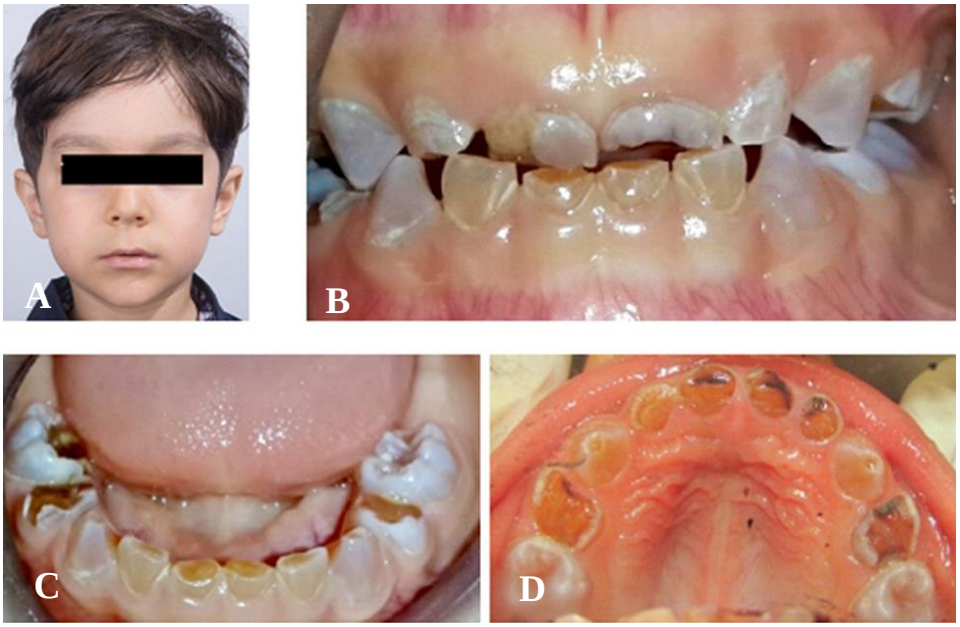


**Fig. 1:** (A) Normal frontal view, (B) Intraoral frontal view, (C) Mandibular arch, (D) Maxillary arch

The objectives of treatment for children with DI are to preserve vitality, structure and size of the dentition, to provide acceptable esthetics and functional dentition, which in turn minimize nutritional deficiencies and psychosocial distress, and finally to preserve the normal vertical dimension and to allow normal growth of the jaw bones [9,10,12]. The purpose of this case report was to describe the characteristics of a patient with DI and to discuss the treatment plan.

## CASE REPORT

*Clinical examination:* A three-and-a-half-year-old male patient presented to the Pediatric Department of Dental School, Isfahan University of Medical Sciences complaining of brown discoloration of his teeth and excessive decay of his primary teeth. The patient also complained of intermittent pain in the lower right posterior region for the past one month. His primary dentition was grayish brown in color and had a translucent hue. Teeth were small in size and showed excessive decay, with complete loss of enamel in most teeth. Coronal height of the teeth was reduced to one-third of the normal. Exposed dentin was visible in incisors, canines, first molars, and the right mandibular second molar. General appearance of the patient was otherwise normal (Fig. 1).

*Radiographic examination:* Panoramic radiographs revealed partial or complete obliteration of the pulp chamber and root canals of the deciduous teeth, spike-like roots with no periapical pathology, and bulbous crowns. Cervical areas were more constricted than those of normal teeth (Fig. 2).

*Family history:* The patient's family history revealed that his 13 year-old sister had similar dental problems in her primary dentition. Clinical appearance of her permanent teeth was normal but radiographic examination revealed periapical radiolucencies, bulbous crowns, obliteration of pulp chambers and lack of pulp horns (Fig. 3). Parents had no consanguineous relationship.

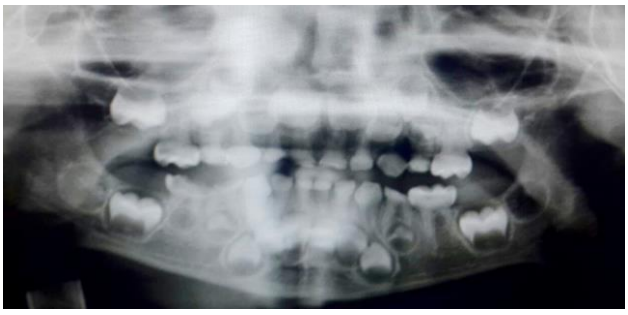

**Fig. 2:** Panoramic radiograph of the patient

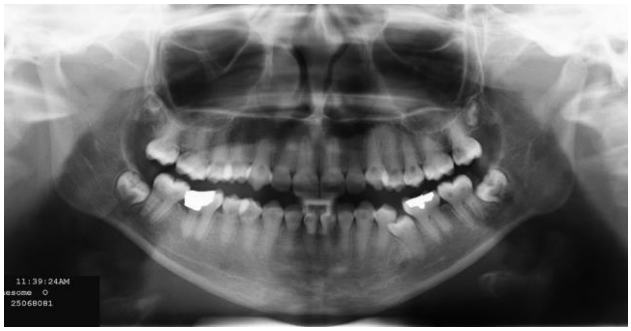

**Fig. 3:** Panoramic radiograph of the patient's sister

On further questioning, the patient's father reported that him, his sister, and brothers suffered from the same condition. There was no history of any unusual bone brittleness or unexplained hearing loss in the family, or any other systemic illness or drug use in the past or the present.

*Treatment plan:* Pulpotomy with mineral trioxide aggregate (ProRoot, Dentsply, Philadelphia, USA) and placement of stainless steel crown (SSC; 3M ESPE, St. Paul, MN, USA) were performed for the mandibular right second molar. After amputating the coronal pulp and controlling hemorrhage, mineral trioxide aggregate was applied with a moistened cotton pellet over. The chamber was sealed with Zonalin (Kemdent Works, Purton, UK). Several days later, during the second appointment, the patient's teeth were asymptomatic. After isolation, the pellet was removed and Zonalin was placed over mineral trioxide aggregate. Due to the very small size of this tooth, a number 2 SSC was cut on the lingual side and was soldered and cemented with polycarboxylate cement (Poly F, Dentsply, DeTrey, Konstanz, Germany) (Fig. 4).

The maxillary and mandibular left molars were restored with number 2 SSC (Figs. 4 and 5). The mandibular right first molar was extracted because of fracture and decay at the gingival level (Fig. 5). The band and loop was inserted during the next visit to maintain the space of this tooth (Fig. 5). The anterior teeth were not restored because the patient was not cooperative and there was no history of pain or abscess. Finally, fluoride therapy was administered, and the patient was discharged home with oral hygiene instructions, and follow-up visits were planned.

## DISCUSSION

Abnormal dentinal structure might be the primary reason for enamel fracture [9,14], resulting in lower numbers, varied diameters, short, twisted, and irregularly distributed dentinal tubules, leading to excessive water content and decreased calcification of dentin [5,8,15].

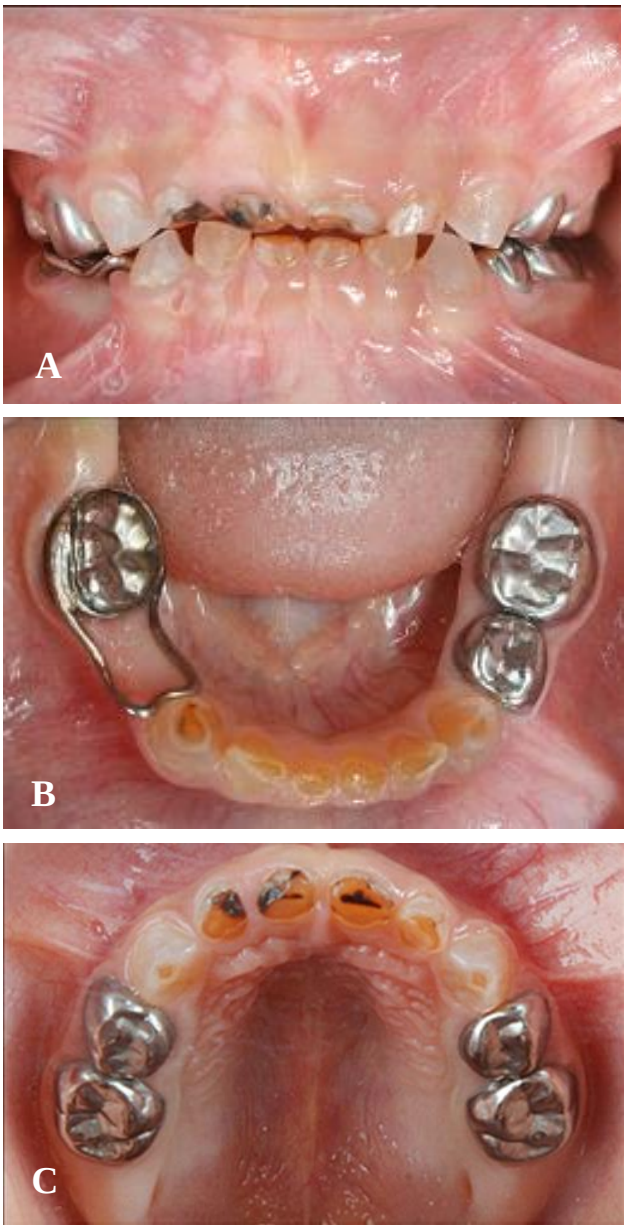


**Fig. 4:** After treatment. (A) Frontal view; (B) The mandibular arch; (C) The maxillary arch

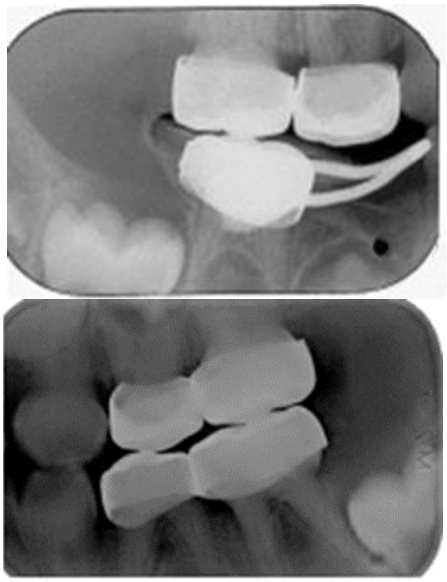

**Fig. 5:** Radiographic examination after treatment

Formerly a defective dentinoenamel junction was described as the reason for susceptibility of DI-affected teeth to excessive decay [8]. However, Levin et al, [16] detected excessive decay with normal scalloping of dentinoenamel junction. In addition, scanning electron microscopic studies in patients with DI have revealed a near normal dentinoenamel junction, somewhat abnormal mantle dentin, and significantly abnormal secondary dentin [8]. It has been concluded that enamel disintegration is mainly caused by abnormal collagen or matrix components formed by defective odontoblasts [8,14]. The patient's age and penetrance of the genetic defect might be ascribed to the extent of enamel wear [1]. The most important factor, which is beyond the dentists' control, is the time at which the patient presents to clinic for treatment. The possibility of delivering optimal treatment decreases as the patients' age increases [1,11,14]. Majorana et al, [17] declared that yellow-brown discoloration is more prevalent than gray discoloration in DI-affected deciduous teeth. However, the prognosis of gray discoloration is better because of lower decay [17,18]. In order to prevent excessive loss of tooth structure, it is recommended to place SSC on deciduous and young permanent posterior teeth as soon as they erupt [3,19]. In our patient, primary incisors, first molars, and lower right second molar exhibited excessive decay and enamel fractures. Hence, we placed crowns on all primary molars to rehabilitate the lost occlusal height. Pulpotomy and placement of SSC was performed for the mandibular right second molar. Pulp treatment was very complicated as there was no specific pulp chamber. Therefore, prognosis might not be excellent. The importance of follow-up examinations was explained to parents and informed consent was obtained. Due to decreased dentin hardness, the restorations will not be permanent and follow-up visits are necessary [3]. When decay occurs at or below the gingival line, as in this case, tooth extraction is indicated [3,20] and the mandibular right first molar was removed. A band and loop space maintainer was indicated to prevent space loss. It should be noted that this child was uncooperative and his parents did not allow us to treat him under general anesthesia or sedation. Therefore, treatment was delivered using behavior modification and limitation methods such as Papoose Board (Olympic Medical Corp., Seattle, USA) and Molt Mouth Prop (Hu-Friedy, Chicago, IL, USA) and the patient's parents were present. While in theory resin bonding to DI-affected teeth is compromised, it is suggested for primary and young permanent anterior teeth [19]. However, in this case deciduous anterior teeth were not restored because of the patient's uncooperative behavior and because there was no history of pain or abscess [3]. Oral examination of the patient's 13-year-old sister revealed normal appearance of the permanent teeth. This is consistent with reports by Schwartz and Tsipouras [21], who concluded that dental decay in DI was more severe in the primary teeth compared to the permanent teeth.

## CONCLUSION

It can be concluded that children with DI should be evaluated as soon as the teeth erupt in order to restore them with SSCs and prevent loss of tooth

structure. Frequent follow up visits are necessary to restore any new enamel fracture and maintain oral health.

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