Case Report on Early Treatment of Dentinogenesis Imperfecta

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Introduction

Dentinogenesis imperfecta (DI) is an autosomal dominant genetic disorder of type I collagen that affects dentin at histodifferentiation stage during tooth development. It can be present alone (DI Type II or III) or be in association with osteogenesis imperfecta (DI type I)[1,2]. The incidence of DI is about one in 8000[3].

Distinctive features include:

- blue-gray/yellow-brown discoloration of teeth
- enamel fracturing
- bulbous crowns
- cervical constriction
- wear/attrition of the teeth

Two major concerns of DI are excessive wear and loss of vertical dimension of occlusion. Early diagnosis and preventive interventions are critical to avoid the negative social and functional consequences.

Case Description

2.5-years-old male with presented to dental clinic with mother being concerned about patient having discolored teeth. Mother denied past medical history, including bone disorders. Upon questioning, patient's father and younger brother present with the same dental issue.

Clinical and radiographic examination revealed:

- blue-gray discoloration of all primary teeth,
- incisal wear
- enamel fracturing of anterior teeth
- molars bulbous crowns
- cervical constriction

Based on findings, patient was diagnosed with DI type II.

Figure 1:
Initial Clinical Photos

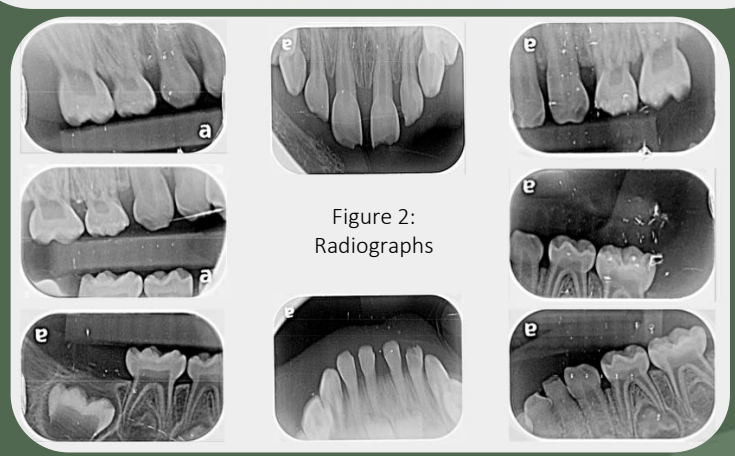

Figure 2:
Radiographs

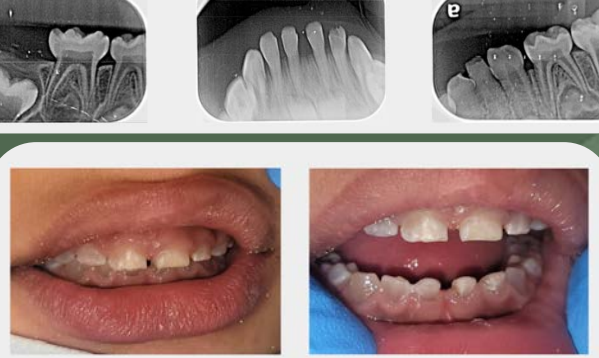

Figure 3: Clinical photos after placement of unfilled resin material on anterior teeth

Discussion

The patient is on the waitlist for comprehensive oral rehabilitation under general anesthesia. The treatment plan includes full coverage restorations (i.e. stainless steel crowns, prefabricated zirconia crowns/resin composite crowns). The rationale behind this treatment is to prevent loss of vertical dimension, provide the patient with a functional and esthetic dentition and maintain dental health and preserve vitality, form, and size of the dentition.

Unfilled resin material was placed on the anterior teeth as an interim solution for preservation of residual enamel to enhance bonding, reinforcement of enamel and dentin and prevention of sensitivity and further attrition of the teeth[4]. Patient is seen for frequent recalls and receives fluoride application until definitive treatment under general anesthesia can be rendered.

There are different potential pathways for prevention and treatment of oral manifestations in the DI population. Depending on severity of enamel fracturing and rapid dental wear the treatment of choice can vary from routine restorative treatment to full coverage restorations in both the primary and permanent dentition[5,6]. Overdenture therapy may be considered in extreme situations. Treatment recommendations should be tailored depending on severity of each case.

References

1. American Academy of Pediatric Dentistry. Guideline on dental management of heritable dental developmental anomalies. Pediatr Dent. 2013;35(5):E179-E184.
2. Neville BW, Damm DD, Allen CM, Bouquot JE. Abnormalities of Teeth. In: Oral & Maxillofacial Pathology. 3rd ed. Philadelphia, Pa: WB Saunders Company; 2009:99-112.
3. Witkop CJ. Hereditary defects in enamel and dentin. Acta Genet Stat Med 1957;7(1):236-9.
4. Massé L, Etienne O, Noirrit-Esclassan E, Bailleul-Forestier I, Garot E. Dentine disorders and adhesive treatments: A systematic review. J Dent. 2021;109:103654.
5. Sapir S, Shapira J. Dentinogenesis imperfecta: An early treatment strategy. Pediatr Dent 2001;23(3):232-7.
6. Sapir S, Shapira J. Clinical solutions for developmental defects of enamel and dentin in children. Pediatr Dent 2007;29(4):330-6.