Two Siblings with Dentinogenesis Imperfecta Type II: Case Reports

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Introduction

Dentinogenesis Imperfecta (DI), hereditary opalescent dentin, affects both primary and permanent teeth through its mesodermal dysplasia lineage. It is an autosomal dominant trait, characterized by robust penetrance and minimal mutation occurrences. [1, 2] DI manifests in three forms: Type 1, associated with osteogenesis imperfecta [3, 4]; Type 2, similar to Type 1 but lacking the osteogenic component; and Type 3, a rare variant found exclusively in Maryland's diverse Brandywine community.[5] This study explores the objectives and nuanced approaches to treating DGI Type II within a family, emphasizing the challenges of dental care.

Case Report

These two patients came for dental examination and were advised to undergo treatment under general anesthesia (GA) due to behavioral problems, including significant communication difficulties and severe dental caries. The parents reported that the children experienced chewing difficulty, refused usual food intake, and cried at night due to toothache.

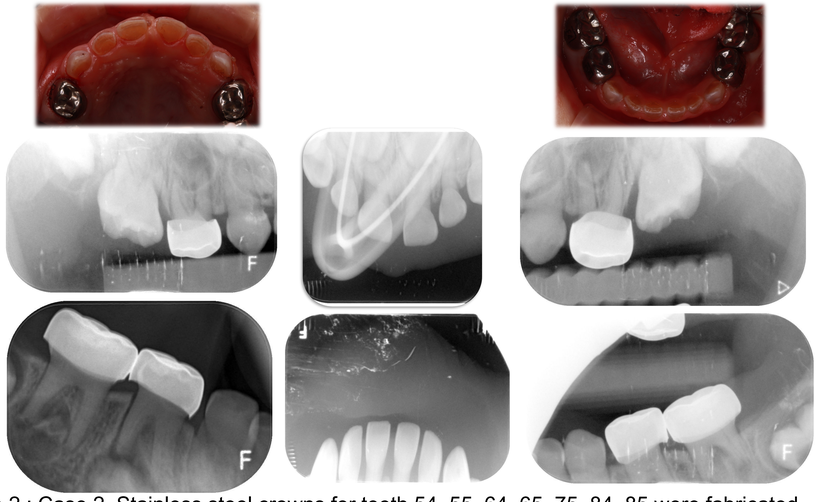
- Case 1: A 3-year-old patient, intraoral examination revealed multiple dental caries in teeth 54, 64, 74, 75, 84, 85.
- Case 2: A 3-year-old patient, intraoral examination revealed multiple dental caries in teeth 54, 55, 64, 65, 74, 75, 83, 84, 85.

Treatment Plan

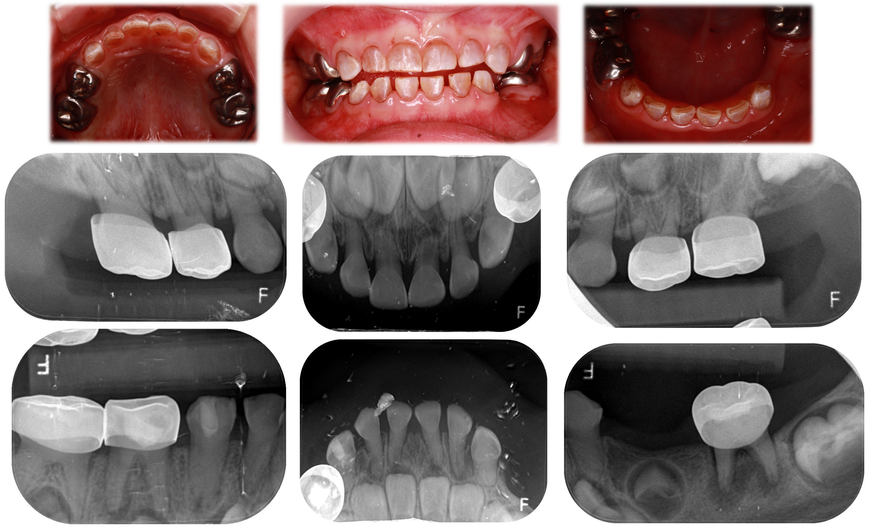
The treatment plan involved general anesthesia with nasoendotracheal tube (NETT) intubation, followed by a routine aseptic procedure. Stainless steel crowns were meticulously fabricated and cemented using Fuji One glass ionomer cement. The primary dentition tends to be more severely affected than permanent dentition, as evidenced by rapid wear and increased caries susceptibility. In early primary dentition, protecting primary molars with stainless steel crowns shortly after eruption is essential. The treatment strategy aims to maintain dental health and preserve the vitality, form, and size of the dentition, provide a young patient with an aesthetically pleasing appearance to prevent psychological issues, and ensure functional dentition.

Clinical and Radiographic Features

- Figure 1 : Case 1. Stainless steel crowns for teeth 54, 64, 74, 75, 84, 85 were fabricated and cemented with Fuji One glass ionomer cement.



- Figure 2 : Case 2. Stainless steel crowns for teeth 54, 55, 64, 65, 75, 84, 85 were fabricated and cemented with Fuji One glass ionomer cement. 83 OD. Extraction of deep caries over 74.



Discussion

Primary teeth affected by DGI-II often show thin enamel and early dentin exposure, leading to rapid and extensive attrition. This necessitates an early diagnostic protocol and comprehensive management strategies. Optimal long-term treatment includes full mouth dental rehabilitation with stainless steel crowns (SSC) and aesthetic prefabricated crowns. Early diagnosis and parental education about DGI are crucial to prepare for the varied dental challenges as children transition to permanent dentition.

Conclusion

This study examines siblings from the same family affected by Dentinogenesis Imperfecta Type II (DGI Type II), an autosomal dominant disorder.[2] The occurrence of DGI Type II in both brothers and sisters highlights the disorder's genetic consistency. The symptomatic treatment with stainless steel crowns for primary teeth is pivotal in restoring oral function and managing DGI Type II effectively.

References

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