Dentinogenesis Imperfecta: A Case Report

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BACKGROUND

Dentinogenesis imperfecta (DI) is an inheritable dental defect originating during the histodifferentiation stage of tooth development. It involves a defect within the predentin matrix that results in amorphic, disorganized, and atubular circumpulpal dentin. There are three types of DI (Shields Types I, II, and III) which present with slightly different manifestations. Radiographs are required to diagnosis this condition, along with a family history and clinical examination. Severity varies considerably between patients, and the appearance of primary dentition does not reliably predict the appearance of the permanent dentition. The degree of expressivity is variable even within the same individual.

DI affects about 1 in 8,000 people, with Shields Type II being the most common. Shields Type III was first identified in a triracial group in Maryland (the Brandywine population) and can also affect persons of Ashkenazi Jewish heritage.

ETIOLOGY

DI is inherited in an autosomal dominant pattern. Type I occurs as part of osteogenesis imperfecta (OI), which is most often caused by mutations on COL1A1 or COL1A2. Type II and Type III are both due to a mutation of DSPP on chromosome 4q21.3.

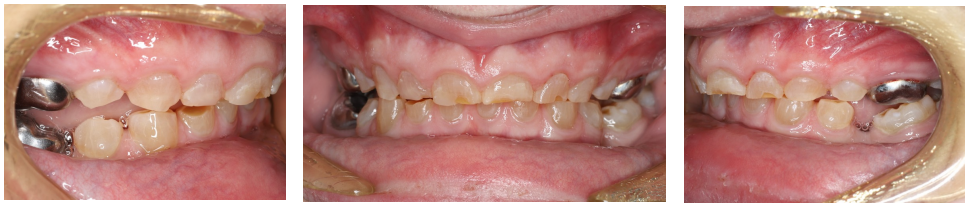
SYMPTOMS

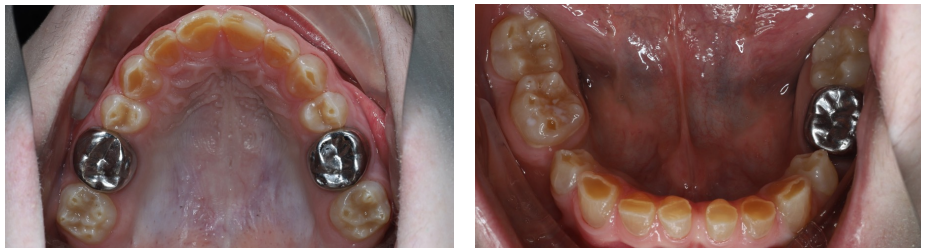
General manifestations include normal intelligence and generally good health unless coupled with OI. Both primary and permanent dentitions are typically affected and are susceptible to extreme wear.

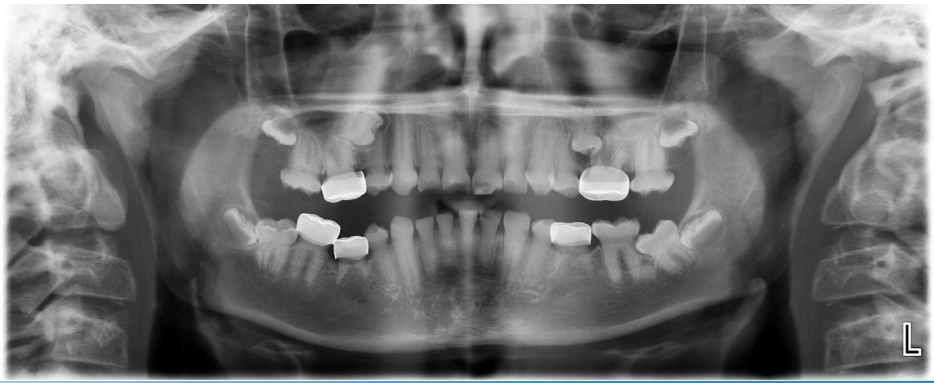
Type I: amber tooth color, bulbous crowns, obliteration of pulp chambers, primary teeth tend to be more affected than permanent teeth, the least severe type of DI, occurs with OI
Type II: same presentation as Type I, the most common type, primary and permanent dentitions tend to be equally affected
Type III: shell-like appearance of teeth with an opalescent hue, enamel pitting, short roots, enlarged pulp chambers, predominance of bell-shaped crowns especially in the permanent dentition, most severe type

Some researchers believe Type II, Type III, and dentin dysplasia Type II are different forms of a single disorder.

CLINICAL PRESENTATION







HISTORY

15 year, 1 month male
Health History: Dentinogenesis imperfecta Type II, otherwise healthy
Medications: cetirizine prn; no known drug allergies
Family History: none recorded; however, dad reports several family members have "the same kind of teeth"
Dental History: DI, congenitally missing #20, #29
Treatment since pan: Extraction of #1, #4, #13, #16, #17, #K, and #T, #32; luxation and surgical exposure of #18 and #31

Patient will complete Phase II orthodontics, followed by jaw surgery to correct his Class II skeletal discrepancy.

CONSIDERATIONS

Stainless steel crowns (SSCs) can be used to prevent excessive wear on primary molars and should be placed as soon as wear is apparent. Abscessed primary teeth require pulp therapy, but extraction is indicated if pulpal obliteration is present. As adults, most patients require full-coverage crowns and frequent root canal therapy.

If esthetics of the primary dentition becomes a concern, anterior pre-veneered SSCs or partial overdentures can be used. In the permanent dentition, tray bleaching can be used to lighten the shade of teeth, then composite or porcelain veneers can be placed. Full-coverage crowns on permanent teeth may also be used to improve esthetics, help prevent wear, and maintain the vertical dimension of occlusion.

Every effort should be made to maintain the teeth as long as possible to maximize the options the individual has as an adult.

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