

# Dentin Dysplasia: A Rare Genetic Disorder Impacting Tooth Development and Structure

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## DESCRIPTION

Dentin dysplasia is a rare genetic disorder that affects dentin, a mineralized tissue that forms the bulk of teeth. It is characterized by disturbances in dentin formation, leading to abnormal tooth development and structural abnormalities.

Dentin dysplasia is classified into two types: Type I, also known as radicular dentin dysplasia, and Type II, known as coronal dentin dysplasia. This study aims to provide a comprehensive study of dentin dysplasia, including its etiology, clinical features, radiographic findings, and management strategies.

## Etiology

Dentin dysplasia is an autosomal dominant disorder, which means that an affected individual has a 50% chance of passing on the condition to each of their children. Mutations in the Dentin Sialophosphoprotein (*DSPP*) gene have been identified as the underlying cause of dentin dysplasia. The *DSPP* gene is responsible for the production of dentin matrix proteins critical for normal tooth development.

## Clinical Features

**Type I dentin dysplasia (radicular dentin dysplasia):** In Type I dentin dysplasia, the root portion of the tooth is predominantly affected. The clinical features include short and blunted roots, periapical radiolucencies, and premature loss of primary and permanent teeth. The teeth may appear normal in shape and color, but they are prone to fractures and have a high risk of pulp infections and abscesses.

**Type II dentin dysplasia (coronal dentin dysplasia):** Type II dentin dysplasia primarily affects the coronal dentin, the part of the tooth above the gum line. The clinical features include bulbous crowns, amber discoloration, and marked attrition. The teeth may also exhibit shell-like appearance due to defective dentin formation. The roots are usually unaffected, and patients may experience spontaneous tooth loss or require extraction due to extensive coronal breakdown.

## Radiographic findings

Radiographic examination plays a crucial role in diagnosing dentin dysplasia. In Type I dentin dysplasia, the roots are short and blunt, with apical radiolucencies that may mimic periapical pathology. In Type II dentin dysplasia, the pulpal chambers are obliterated, resulting in a "thistle tube" or "flame-shaped" appearance on radiographs.

## Management

The management of dentin dysplasia focuses on preventing complications and maintaining oral health. It requires a multidisciplinary approach involving dentists, orthodontists, and oral surgeons. The treatment options may include:

**Regular dental care:** Patients with dentin dysplasia require meticulous oral hygiene practices and regular dental check-ups to prevent dental caries, gum disease, and infections. Fluoride treatments and sealants may be recommended to strengthen and protect the tooth structure.

**Restorative dentistry:** Restorative procedures, such as composite resin or porcelain crowns, may be necessary to protect and restore the weakened teeth. Dental professionals should carefully evaluate the structural integrity of the affected teeth before performing any restorative work.

**Prosthetic replacement:** In cases where teeth are extensively damaged or lost, prosthetic replacements such as dentures, bridges, or dental implants may be considered to restore aesthetics and function.

**Orthodontic intervention:** Orthodontic treatment may be required to correct any misalignment or malocclusion caused by the abnormal tooth structure. Braces or other orthodontic appliances can help align the teeth and improve the overall bite.

**Genetic counseling:** Genetic counseling should be offered to individuals and families affected by dentin dysplasia to provide information on the inheritance pattern, risk of recurrence, and available reproductive options.

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**Received:** 19-Jun-2023, Manuscript No. JOY-23-25561; **Editor assigned:** 22-Jun-2023, PreQC No: JOY-23-25561 (PQ); **Reviewed:** 07-Jul-2023, QC No. JOY-23-25561; **Revised:** 14-Jul-2023, Manuscript No: JOY-23-25561 (R); **Published:** 21-Jul-2023, DOI: 10.35248/JOY.23.7.673

**Citation:** Eduardo C (2023) Dentin Dysplasia: A Rare Genetic Disorder Impacting Tooth Development and Structure. J Odontol. 7:673.

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Dentin dysplasia is a rare genetic disorder characterized by disturbances in dentin formation, leading to structural abnormalities and dental complications. Early diagnosis, comprehensive dental care, and a multidisciplinary approach are essential for managing the condition and improving the quality

of life for affected individuals. Further analysis and genetic studies are needed to better understand the underlying mechanisms of dentin dysplasia, which will contribute to improved diagnosis, treatment, and genetic counselling in the future.