



## Endodontic Management of Dentin Dysplasia Type II in a Pediatric Patient: A Case Report

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Dentin Dysplasia Type II (DD-II) is a rare genetic disorder that affects the coronal dentin, often leading to pulp stones and complicated endodontic treatment. This case report presents the endodontic management of DD-II in an 11-year-old female patient. The patient, referred for root canal treatment, exhibited symptoms of moderate pain and was diagnosed with pulp necrosis in tooth #19; radiographic findings revealed multiple pulp stones, pulp canal calcification, and apical radiolucency. Endodontic treatment was performed, involving the removal of pulp stones with ultrasonic tips, careful cleaning and shaping of the root canals, and obturation using gutta-percha and AH-26 sealer. Twelve months post-treatment, the patient showed complete healing of the apical lesion, with no related complications. This case underscores the importance of early diagnosis and timely endodontic intervention for favorable outcomes in patients with DD-II.

**Keywords:** Dental Pulp Calcification; Dentin Dysplasia; Endodontic; Genetic Diseases; Multidisciplinary Approach; Pediatric Dentistry; Root Canal Therapy

### Introduction

Dentin Dysplasia (DD) is a rare hereditary disorder [1] that affects dentine. It is classified into two main types: Dentin Dysplasia Type I (radicular dentin dysplasia) and Dentin Dysplasia Type II (coronal dentin dysplasia) [1-4]. Both types are inherited in an autosomal dominant pattern [1, 5], meaning a single copy of the mutated gene (from one parent) is sufficient to cause the disorder. While Dentin Dysplasia Type I primarily affects the root structure of the teeth [6], leading to shortened or absent roots, Dentin Dysplasia Type II (DD-II) primarily affects the coronal portion of the tooth, resulting in characteristic pulp abnormalities, including the presence of pulp stones [7].

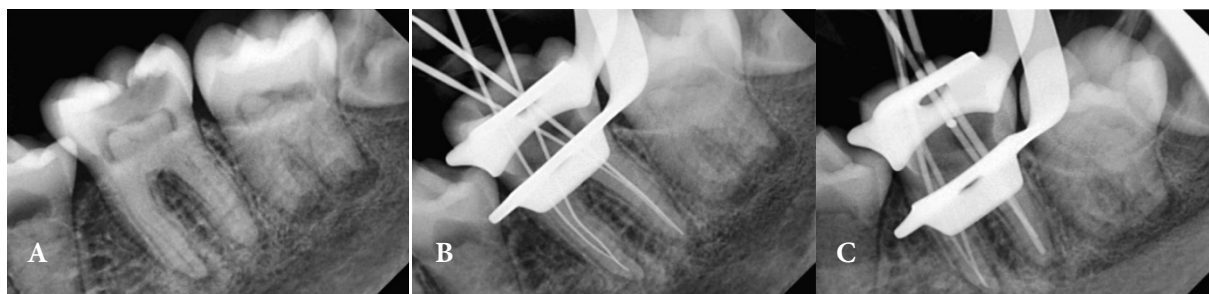
DD-II is less common than Type I (1 in 100,000 for Type I) [8] and is typically diagnosed based on clinical and radiographic findings. In DD-II, the deciduous teeth often exhibit yellow-brown or amber discoloration, resembling the appearance of dentinogenesis imperfecta. However, in the permanent dentition, the teeth usually appear clinically normal, although

radiographs reveal multiple pulp stones that can obstruct the root canal system and complicate dental treatment. The pulp chambers in the permanent teeth are often thistle-tube-shaped, and the roots tend to have normal morphology, unlike in DD-I, where root development is severely affected [8].

The etiology of DD-II is linked to mutations in the DSPP gene (dentin sialophosphoprotein) [9], which plays a crucial role in the formation of dentin [4]. Mutations in this gene lead to the formation of abnormal dentin and pulp stones. These pulp stones can cause pulpal necrosis, even in the absence of trauma or caries, and may result in periapical pathology [10]. The clinical management of DD-II can be challenging due to the presence of pulp stones, which complicate endodontic treatment [11].

In this paper, we report a case of DD-II diagnosed during a routine root canal treatment of a permanent tooth in an 11-year-old female patient. We describe the specific radiographic images of the patient's mixed dentition and the particularities that influence the differential diagnosis and treatment outcomes as well. To illustrate these characteristics and management challenges, we present a case of DD-II diagnosed during routine dental treatment.





**Figure 1.** A) Initial radiography of the patient; B) Working length determination; C) Master apical cone radiography



**Figure 2.** A) Post obturation radiograph; B) 12-month follow up

## Case Presentation

An 11-year-old girl without any systemic diseases, allergic reactions, or regular drug consumption was presented to the pediatric dental department with a referral to the endodontic department for root canal treatment. The patient reported experiencing occasional pain in her back teeth and moderate, intermittent pain on the left side of the mandible. However, no facial swelling or lymph node involvement was detected during the extraoral examination. The intraoral examination revealed no signs of a sinus tract, swelling, or discoloration. However, the mandibular first molar [12] exhibited caries, and the mandibular right second premolar had not yet erupted. Results of the pulp diagnostic tests (Table 1).

The pulpal diagnosis indicated infected pulp, while the periapical diagnosis was chronic apical periodontitis. Radiographic findings included occlusal caries, pulp stones, an enlarged periodontal ligament (PDL), and apical radiolucency (Figure 1A).

Written informed consent was obtained from the patient and is documented under the approval number IR.ZAUMS.REC.1403.338. According to the treatment plan, after obtaining informed consent, the tooth was anesthetized using an inferior alveolar nerve block injection with 1.8 ml of lidocaine 2% with epinephrine 1:80,000 (Daroupakhsh, Tehran, Iran). After placing a rubber dam, the access cavity was created with a high-speed diamond round bur No. 2 (Jota

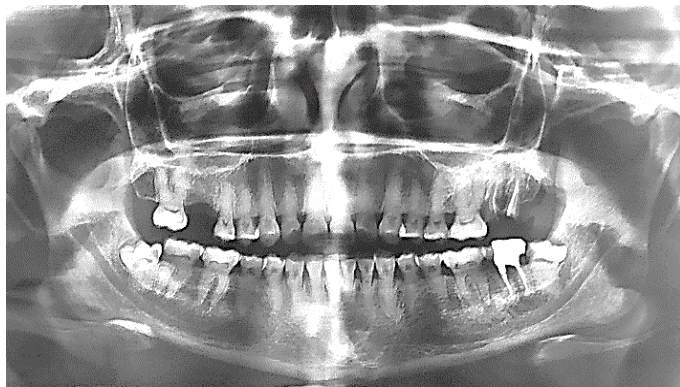
AG, Rüthi, Switzerland) using continuous water spray under a dental microscope (Carl Zeiss, Meditec Inc., Dublin, CA, USA). The access cavity was refined using a diamond fissure bur No. 2 (Jota AG, Rüthi, Switzerland). Pulp stones were carefully removed with an ultrasonic tip (blue tip, Eighteenth, China), and the shape of the pulp chamber was established. The pulp chamber was enlarged with a long-shank Muller bur and irrigated frequently with hypochlorite and saline. Four orifices were located and explored using a #15 C-file (M3 Gold series, Düsseldorf, Germany). Coronal preflaring was performed with an SX instrument. The working length was determined using a Root ZX apex locator and confirmed with a radiograph (Figure 1B).

The initial file used was size 15 for both the mesial and distal canals. The Sp1 Gold rotary system was applied. A rotary file S1 (UDG, Changzhou, China), which reached the working length by a picking motion, was utilized. Subsequently, S2, F1, and F2 files were used respectively to achieve the working length. Canals were irrigated with 2% NaOCl and normal saline during instrumentation. Master cones of size 30/2% for mesial canals and 40/2% for the distal canal were selected. The canals were thoroughly dried with sterile paper points (META, Chugbuk, South Korea), and obturation was performed using Gutta-percha (META, Chugbuk, South Korea) and AH-26 sealer (Dentsply DeTrey, Konstanz, Germany) via the lateral condensation technique. Finally, the access cavity was restored with composite resin (Figure 2).

**Table 1.** Diagnostic pulp tests

| Tooth Number | Cold | Heat | EPT | Percussion          | Palpation | Mobility |
|--------------|------|------|-----|---------------------|-----------|----------|
| #19          | -    | -    | -   | Different sensation | N         | WNL      |
| #14          | N    | -    | +   | N                   | N         | WNL      |
| #13          | N    | -    | +   | N                   | N         | WNL      |
| #3           | N    | -    | +   | N                   | N         | WNL      |

EPT :Electric pulp test; N: Normal; WNL: Within normal limit



**Figure 3.** OPG of patient's mother shows the signs of DD-II

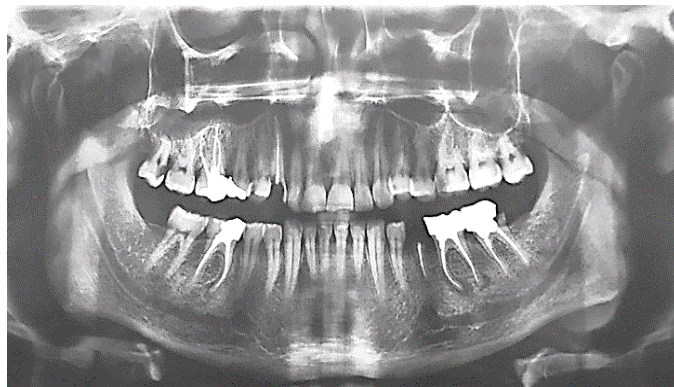
The patient was followed up for 12 months; however, there were no related complications. Upon taking a detailed family history, it was revealed that the patient's mother had been diagnosed with DD-II, while her father was unaffected. This information suggested a potential genetic inheritance of the condition from the mother. The patient had no known systemic diseases, allergic reactions, or regular drug consumption (Figures 3, 4).

## Discussion

In humans, hereditary dentin defects are generally divided into two main categories based on their physical characteristics: dentinogenesis imperfecta and dentin dysplasia. Dentin dysplasia is further split into two types: type I and type II, with type II being the less severe form [13]. DD-II is a rare genetic disorder affecting dentin formation. It is associated with genotyping for microsatellite alleles specific to the area of chromosome 4q linked to DGII, and the similarity of the primary dentition phenotype suggests the involvement of the gene for dentin dysplasia [14, 15].

This case report presents the successful management of DD-II in an 11-year-old patient, highlighting the unique challenges and considerations in treating this rare genetic disorder. This case is notable due to the rarity of this genetic condition [13, 16, 17]. The presence of pulp stones [18] and periapical radiolucency in the absence of significant caries [8] or trauma highlights the unique diagnostic and treatment challenges associated with this condition.

In this case, the presence of pulp stones and chronic apical periodontitis in tooth #19 aligns with the characteristics of this condition. The successful management of this case underscores the importance of tailored treatment approaches for rare dental anomalies. The prognosis is favorable, given the success of the root canal therapy and the absence of complications.



**Figure 4.** No sign of DD-II in patient's father

Building on the clinical presentation, it is important to note that DD-II typically follows an autosomal dominant inheritance pattern. This pattern is evident in our patient, who had a family history of the condition. The clinical presentation in our patient aligns with the characteristic features of DD-II, including a normal enamel appearance but with underlying dentin abnormalities. Radiographically, we observed the typical “thistle tube” appearance of the pulp chamber, with a sudden constriction at the base and very thin root canals. This is consistent with the descriptions provided by previous case reports and reviews.

DD-II typically follows an autosomal dominant inheritance pattern. Consistent with previous reports demonstrating a clear hereditary pattern, our patient had a family history of the condition, as her mother was affected. This aligns with reports by Paula Perlea *et al.* [13] and Deepak Daryani *et al.* [19], which documented DD-II affecting multiple family members across generations. Additionally, our patient lacked any systemic disorders, whereas some prior cases have noted associated conditions. For example, Daryani *et al.* [19] reported DD-II in siblings with short stature, hypocalcemia, and other systemic findings. However, it is worth noting that rare cases of autosomal recessive inheritance have been documented, highlighting the genetic complexity of this disorder.

Although the exact etiology of DD-II remains unclear, several theories have been proposed. Logan *et al.* [12] suggested that degenerative changes in dental papillae lead to delayed growth, abnormal calcification, and pulp chamber obliteration. Wesley *et al.* [20] proposed that DD results from abnormal odontoblast differentiation or function due to atypical interactions with ameloblasts. Additionally, ectopic dentin formation may occur due to early radicular epithelial sheath ingrowth or the displacement and proliferation of internal dental organ cells in the dental papilla [21].

Previous reports reviewed other signs that were not found in our case, including microdontia among permanent dentition, partial ankyloglossia, early exfoliation of deciduous teeth, and loss of tooth structure in permanent teeth [19]. They also pointed out skeletal disorders, which are not evident in this report. Despite this, almost all of them reported pulp stones and the "thistle tube" appearance collectively.

This case highlights the need for further research into the genetic basis of DD-II and its potential associations with systemic conditions. Long-term follow-up studies on patients with DD-II would be valuable for understanding the progression of the condition and the efficacy of different treatment approaches over time.

## Conclusion

The successful management of the presented case underscores the necessity for early diagnosis and intervention in patients with DD-II. This report highlights the importance of recognizing and managing DD-II. Early diagnosis and intervention are critical for preserving affected teeth and preventing severe complications. The complexity of this disorder necessitates a multidisciplinary approach in both diagnosis and management.

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## Conflict of interest

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## Authors' contributions

Conceptualization: ES/SR; Methodology: ES/SR; Formal Analysis and Investigation: ES/SR; Writing-Original draft preparation: ES/SR; Writing-review and editing: ES/SR; Supervision: ES. All authors read and approved the final manuscript.

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