Endodontic Treatment of Dentin Dysplasia Type I D

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Abstract

Dentin Dysplasia (DD) Type I is a developmental condition affecting dentin, inherited in an autosomal dominant pattern or occurring due to a new mutation. While the crowns of DD Type I affected teeth appear clinically normal, the roots are blunt and shortened. Pulp necrosis and periapical pathoses may be seen in the absence of obvious causes. Pulp stones and calcifications are frequently encountered. Endodontic management of DD may be challenging. A case of DD Type I, sub-classification d, in which spontaneous irreversible pulpitis developed on three mandibular incisors is documented. The case was managed by conventional endodontic treatment. Knowledge of this uncommon dental condition may assist dentists to adequately diagnose and manage these cases. Extraction should not be considered the first-line treatment option when sufficient root length is available to attempt endodontic treatment. Referral for medical evaluation is recommended to rule out systemic diseases which may mimic this condition.

Keywords: dentin dysplasia, endodontics, pulp stones, rootless teeth, shortened roots

Introduction

Dentin dysplasia (DD) was first described as “rootless teeth” by Ballschmiede in 1920 and later formally named by Rushton in 1939. This uncommon dental condition (OMIM #125400) may occur either sporadically or be inherited in an autosomal dominant pattern with complete penetrance (1-3).

DD is considered a rare entity, with a reported prevalence rate of approximately 1:100 000 (4). The condition is characterised by abnormal dentin formation, which may affect both the primary and permanent dentition. Clinically, tooth crowns appear normal and at times the first detectable sign of disease is premature tooth exfoliation due to severe mobility (5). The clinical and radiological presentation of DD is generally classified into two distinct types: DD Type I, also known as the radicular type; and DD Type II, the coronal type (6).

Teeth affected by DD Type I display normal coronal enamel and dentin. The radicular dentin however demonstrates a loss of organisation (4). Amorphous dentin deposition can lead to restricted dentin formation resulting in “rootless” teeth. Alternatively, dentin formation may proceed unchecked, manifesting in the appearance of multiple pulp stones (7). These pulp stones present as dentinal islands consisting of dysplastic dentinal tubules and numerous calcified bodies (8).

Radiographically, the roots of teeth affected by DD Type I appear short and blunted. Multiple periapical radiolucencies may be seen despite the absence of an obvious cause (*e.g.*, extensive caries or deep restorations) (4). Radiographically, DD Type I can be further classified into four subtypes: DDIa, DDIb, DDIc and DDId (Figure 1), according to the degree of dentin disorganisation (3).

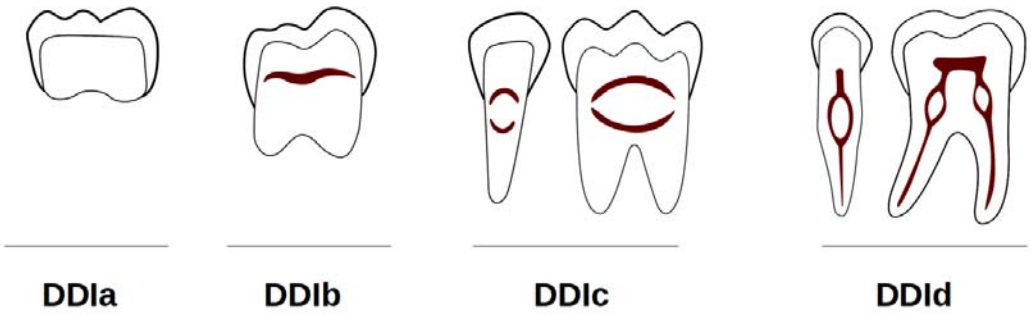

Figure 1. Sub-classification of DD Type I.

DD1a represents a complete lack of root formation with absence of pulp chambers and frequent periapical radiolucencies. DD1b displays significantly shortened roots with one horizontal crescent-shaped pulp chamber. Periapical radiolucencies are also frequently seen. DD1c also shows shortened roots, however, two horizontal crescent-shaped pulp chambers surrounding a central dentinal island are visible in this subtype. This subtype may display variable frequency of periapical radiolucencies. DD1d shows normal root length with pulp stones which are often generalised throughout the patient's dentition. The radiographic appearance of the pulp stones in this subtype is commonly described as a "stream flowing around boulders". Localised bulging of roots may occur around enlarged pulp stones. Few periapical radiolucencies are seen in this subtype (4). Varying subtypes of DD may be present in the same patient.

Although overlapping features may exist, DD Type I and II are considered distinct entities and should not be confused (9). In the primary dentition, DD Type II displays features similar to that seen in dentinogenesis imperfecta (*e.g.*, pulp chamber obliteration, bulbous crowns, cervical constriction and thin roots). Permanent teeth

affected by DD Type II, however, do not share the coronal features of their primary counterparts (7). Radiographically, permanent teeth affected by DD Type II demonstrate pulp chamber enlargement described as a “thistle tube-shaped” appearance, with or without pulp stone formation (4).

The dental management of both DD Type I and II may be challenging. Pulpal necrosis may develop spontaneously in teeth affected by DD due to the presence of dentinal islands resembling pulp stones. These stones may result in decreased pulpal perfusion (10). Whilst extraction was historically recommended as the primary treatment option for abscessed or necrotic DD teeth (11), endodontic treatment approaches have also been attempted in both types of DD with varying outcomes (10,12,13).

The present study documents the diagnosis and management of a patient with DD Type I, sub-classification d (DDId), who developed spontaneous pulpitis on three teeth. The case was successfully managed by means of conventional endodontic treatment and non-surgical endodontic retreatment, which included the aid of modern endodontic treatment adjuncts.

Ethical approval for this study was obtained from the Research Ethics Committee, Faculty of Health Sciences, University of Pretoria (Protocol number: 560/2019).

Case Presentation

A 20-year-old female was referred to the Pretoria Oral and Dental Hospital by a general dentist for the treatment of severe pain in the right anterior mandibular area. The right mandibular canine had been extracted due to an inability to achieve endodontic access. Persistent pain after extraction of the canine prompted the referral as the referring dentist was hesitant to continue with further treatment.

Consultation at the Division of Endodontics, Department of Odontology, University of Pretoria revealed no significant findings in the medical history. The past dental history included the provision of several restorations. Clinical examination revealed what appeared to be a normal healthy dentition. No mobility or discoloration of any teeth was noted. Sensibility testing of lower right lateral incisor exhibited severe, lingering pain to cold (Pulpofluorane, Septodont, Saint-Maur-des-Fosses, France). Pulp sensibility testing of the lower right central incisor responded within normal limits. None of the mandibular incisor teeth demonstrated tenderness to percussion. No caries or restorations were present on these teeth.

Periapical and panoramic radiographs were acquired, revealing several absent teeth (Figure 2 and 3). Generalised pulp stones could be seen in almost all the permanent teeth, with the exception of the third molars. Mild taurodontism was also visible in the maxillary third molars.

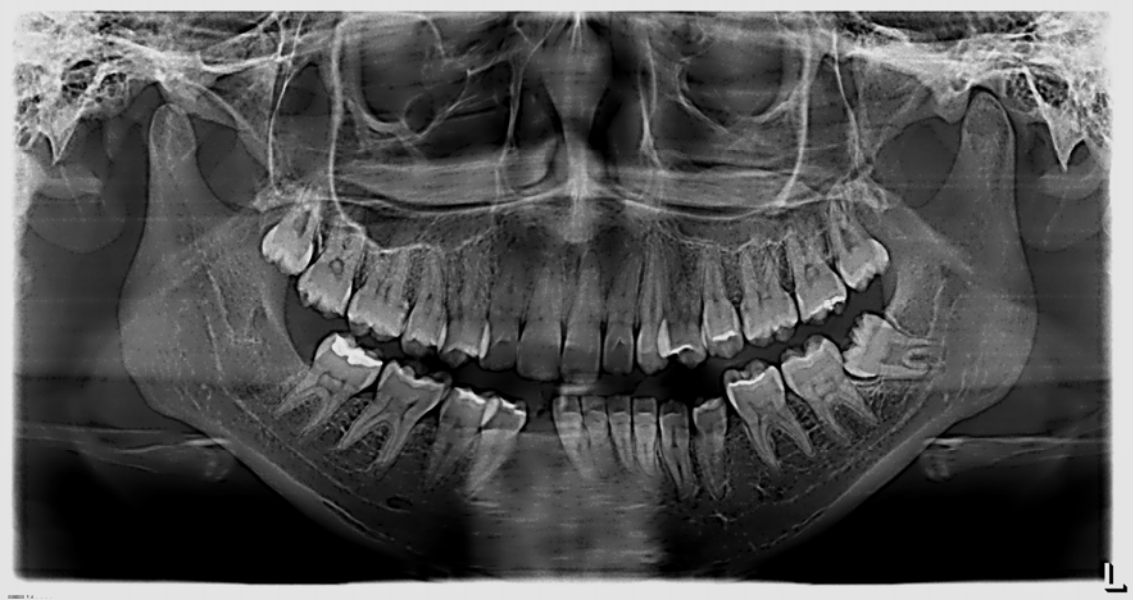

Figure 2. Panoramic radiograph demonstrating generalised pulp stones and absent teeth.

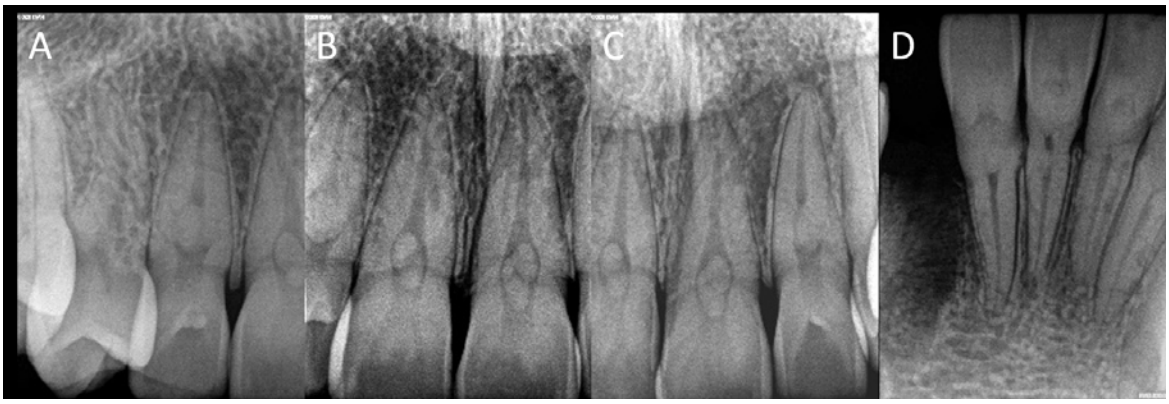

Figure 3. Characteristic “stream flowing around boulders” appearance of the maxillary incisors. Calcifications seen in the root canal systems of the lower right central and lateral incisors. Note the absence of caries and restorations on the mandibular central and lateral incisors.

Large calcifications and/or pulp stones, the absence of caries and minor widening of the periodontal ligament space could be seen at the apices of the lower right central and lateral incisors (Figure 3D). A characteristic “stream flowing around boulders” radiographic appearance with root bulging and pulp stones was seen in the maxillary incisor teeth (Figure 3A, 3B, 3C).

Clinical and radiographic findings suggested a diagnosis of DD Type I, sub-classification d (DDId), with irreversible pulpitis of the lower right lateral incisor. No family history of this condition was reported.

The treatment plan, after obtaining informed consent, included conventional endodontic therapy for the lower right lateral incisor and replacement of the extracted canine with a dental implant. Following local infiltration with lidocaine (Xylotox E80A, 2% lidocaine with epinephrine, Adcock Ingram, Johannesburg, South Africa), isolation

with rubberdam was established. Access to the coronal pulp was achieved through the use of diamond burs.

The canal system at the level of the pulp stone was found to be completely inaccessible due to the presence of amorphous dentin. With the aid of a dental operating microscope (Global, Global Surgical Corporation, St Louis, MO), long-shank Mueller burs (Brasseler, GA, USA), ultrasonic endodontic tips (Start X, Dentsply Maillefer, Switzerland) and C+ files (Dentsply Maillefer, Switzerland), the canal obstruction was successfully negotiated. Access to the apical portion of the canal was established.

Reciprocating (WaveOne Gold, Dentsply Maillefer, Switzerland) and rotary (ProTaper Gold F3, Dentsply Maillefer, Switzerland) instruments were used to shape the canal. Copious irrigation with the use of 3.5% sodium hypochlorite (Jik, Reckitt Benckiser, South Africa) and 17% liquid EDTA (Topclear, Dental Discounts, Johannesburg, South Africa) was performed. Obturation was completed using a matching taper gutta percha cone and GuttaFlow Bioseal endodontic sealer (Coltene/Whaledent AG, Altstatten, Switzerland). The hydraulic condensation technique was employed. The access cavity was restored with composite resin (Filtek Supreme XTE, St. Paul, MN).

Following endodontic treatment of the right mandibular lateral incisor, the pain and symptoms resolved completely. However, approximately one month after the initial endodontic treatment, the neighbouring lower right central incisor spontaneously developed irreversible pulpitis. The same technique as previously described was used to treat this tooth. The only exception was the use of a different endodontic sealer (BioRoot RCS, Septodont, Saint-Maur-des-Fosses, France). Two months following the second root canal treatment, the lower left incisor similarly developed spontaneous

irreversible pulpitis and was treated endodontically. A cone beam computed tomography (CBCT) scan was taken for implant planning purposes prior to the third root canal treatment on the lower left lateral incisor. Axial, coronal and sagittal sections of the mandibular incisors affected by DD can been seen in Figure 4.

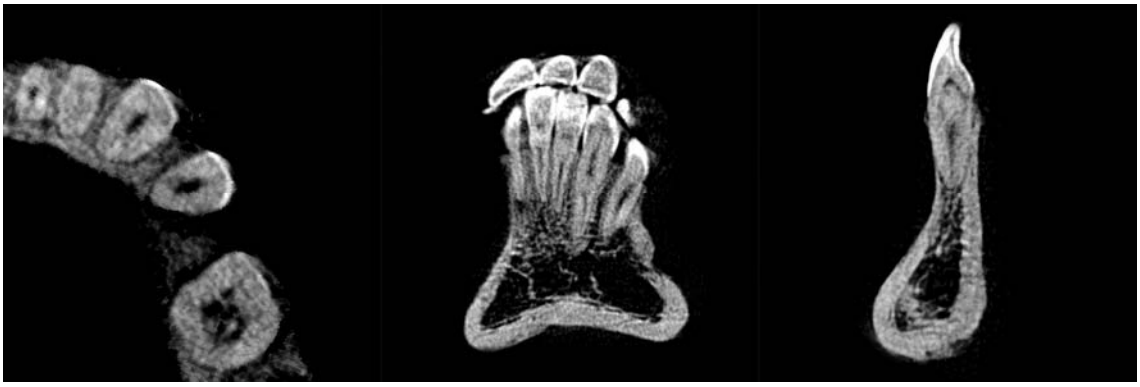

Figure 4. Axial, coronal and sagittal sections of the CBCT scan demonstrating the calcifications seen in the mandibular incisor teeth.

At the 18-month follow-up appointment, complete resolution of the clinical symptoms of all teeth were found. Radiographically, a widening of the periodontal ligament space of the central incisor remained and the decision was made to retreat the tooth non-surgically, despite the absence of any pain or symptoms (Figure 5A and 5B). Long-term follow-up will be required to demonstrate improvement of the radiographic appearance of the apical bone surrounding the central incisor as the healing outcome, form a radiological perspective, remains questionable.

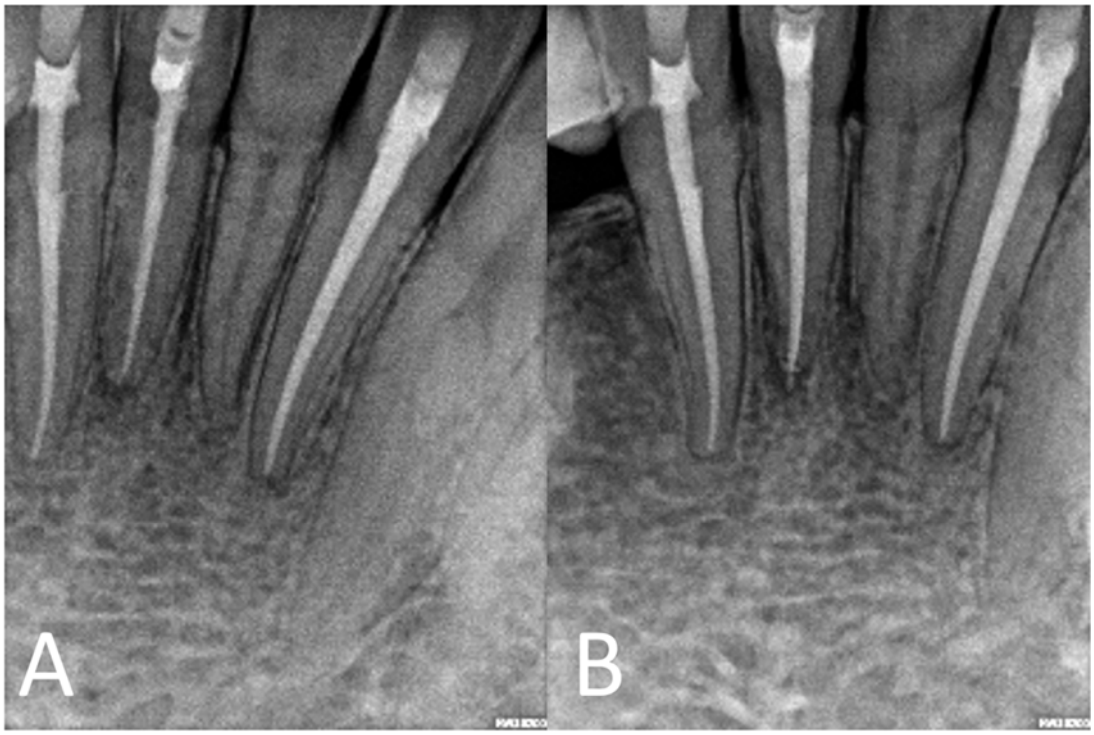

Figure 5. A. Radiographic presentation of the mandibular incisors at the 18-month follow-up visit. **B**. Non-surgical retreatment was initiated on the mandibular right central incisor due to the continued presence of periapical pathosis.

A fibre-reinforced resin bridge was constructed at the six-month visit to restore the extracted canine as a provisional measure to improve aesthetics and maintain space for future implant therapy (Figure 6 – note the normal clinical appearance of teeth affected by DD). The patient was furthermore referred to her medical doctor for physical and haematological investigations to exclude other systemic medical conditions producing calcifications.

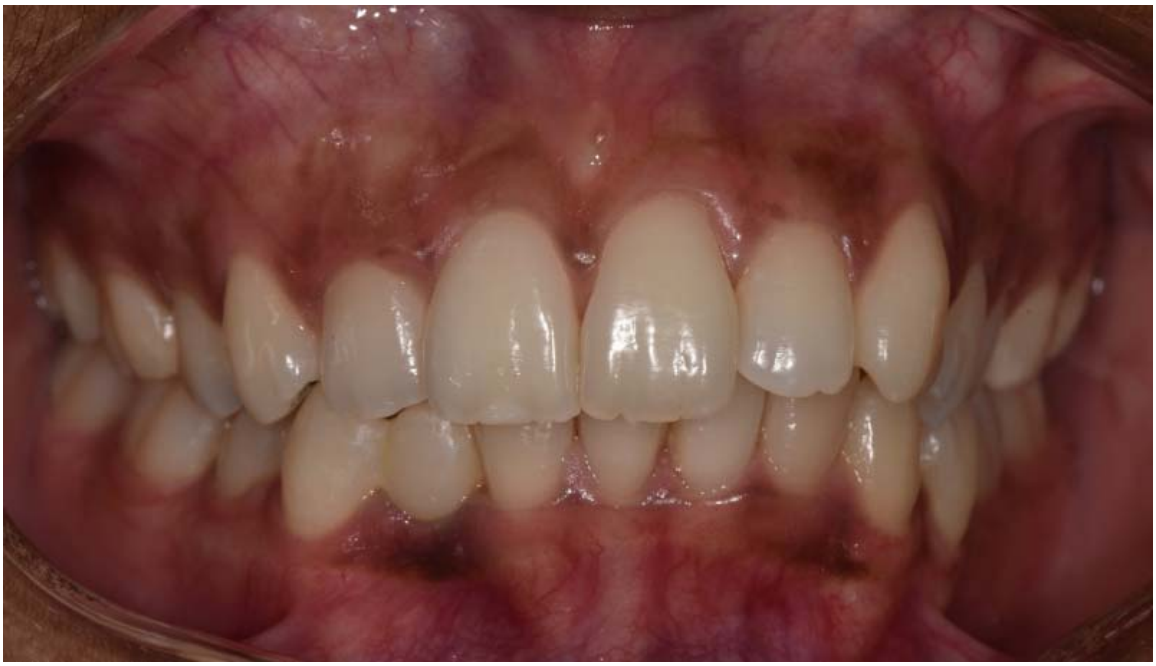

Figure 6. A fibre-reinforced bridge was constructed as a provisional measure at the 6-month follow-up visit to restore the extracted right mandibular canine. Note the clinical presentation of teeth affected by DD appear normal with regards to size, shape and colour.

Discussion

The exact mechanism of development of DD remains unknown. Previous theories have implicated the existence of multiple degenerative foci in the dental papilla (2), early invagination of the epithelial root sheath (14) and abnormal ameloblast-odontoblast interactions (15).

Whilst the molecular basis for the development of DD remains unclear, Xiong and colleagues recently suggested that a mutation in the SSUH2 gene on 3p26.1 may be responsible for the development of autosomal-dominant DD Type I (16). Mutations in the VPS4B and SMOC2 genes have additionally been implicated in the development

of the condition (16-18). Mutation of the DSPP gene in the autosomal dominant forms of coronal dentin dysplasia (Type II) as well as the related entity dentinogenesis imperfecta (19-21) have previously been reported. Not all genes responsible for the development of DD may have been identified (22).

Radicular development is induced and controlled by Hertwig's epithelial root sheath (HERS). It is hypothesised that the disruption of root formation in DD Type I does not stem from the apical growth of HERS but rather a cessation of radicular dentinogenesis. The termination of the process is thought to be caused by odontoblast obstruction by the calcified bodies filling the pulp chamber. The reasons for the formation of these calcified bodies remain unclear (23).

Ravanshad and Khayat proposed that the spontaneous pulpal necrosis seen in DD may occur as a result of impaired pulp circulation and nourishment, rendering the pulp susceptible to bacteraemia (10). The authors of the present case agree with this assertion. This theory may explain the development of the spontaneous irreversible pulpitis of the lower right central and left lateral incisors in the present case. The possibility of traumatic extraction of the lower right canine may however have contributed to the development of pulpitis on the lower right lateral incisor.

Historically, endodontic treatment approaches were uncommon for cases of DD and symptomatic teeth were often extracted (24). Extraction approaches may however be detrimental, especially in younger individuals, due to the alveolar bone loss which accompanies dental extraction. A number of reports have however described successful endodontic management of DD (8,10,12,13,25,26). Endodontic surgery and retrograde filling has additionally been reported as a treatment option for DD Type

I (9,10,26,27). The present case contributes to the existing body of evidence for the viability of endodontic treatment in the management of DD. Due to the number of variations in the clinical and radiographic presentation of the condition, it is difficult to recommend a standardised treatment approach for these cases.

Steidler recommended the prevention of caries and periodontal disease to retain teeth affected by DD for as long as possible (28). The long-term prognosis is however dependent on factors such as the degree of root shortening and ease of access to root canals in symptomatic teeth. Preventive care, such as oral hygiene education along with debridement of supra- and subgingival deposits is important in preventing early tooth loss due to periodontitis (4). Whilst the authors of the present study agree with preventative approaches, it must be considered that despite good oral hygiene – as demonstrated in the present case – spontaneous pulpal inflammation and subsequent pulp necrosis may still occur in patients affected by DD, even in the absence of any significant caries or periodontal problems.

Practical challenges in the clinical management of teeth affected by DD Type I include calcified pulp chambers, multiple pulp stones, a decreased crown-to-root ratio and, in some instances, multiple periapical radiolucencies (4). These challenges are especially significant when endodontic approaches are considered as the primary treatment option (13). Although the present case displayed several such challenges, predictable management was achieved by means of conventional endodontic treatment and non-surgical retreatment. In milder forms of DD, where sufficient root length is available, conventional endodontic treatment should routinely be considered the first-line treatment option (12). The present case supports this assertion.

The importance of magnification, improved illumination and ultrasonic instrumentation during challenging endodontic treatment cases is well-established (29,30). Successful treatment of the present case would not have been possible without the aid of these endodontic treatment adjuncts.

DDId is an unusual diagnosis; other systemic medical conditions which may mimic the changes seen in DD, such as tumoral calcinosis, should be investigated (4). In line with this recommendation, appropriate referral of the patient in the present case was made to a medical practitioner. The findings from both the physical and haematological investigations revealed nothing abnormal, and provided final confirmation of the diagnosis of DD Type I.

In conclusion, this paper reports an uncommonly encountered case of DDId which developed spontaneous irreversible pulpitis of several mandibular incisors. Predictable treatment outcomes were achieved by means of conventional endodontic treatment on two teeth and a questionable outcome of primary endodontic treatment of the mandibular right central incisor necessitated non-surgical retreatment. The latter will require continued follow up to determine long-term radiographic success. The provision of endodontic treatment in this case was greatly assisted by both magnification and ultrasonic instrumentation. Knowledge of this uncommon developmental condition of dentin may assist general dentists and endodontists in the identification, management and/or appropriate referral of these cases. When sufficient root length is available to attempt endodontic treatment, extraction should never be considered the first-line treatment option for teeth affected by DD. Referral of patients with DD for medical evaluation may be required to rule out other systemic diseases which may mimic this condition.

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