Management and Long-Term Follow-Up of a Case with Different Dental Alterations of Dentin Dysplasia: Case Report

Farklı Dental Bulguları Olan Dentin Displazili Bir Olgunun Tedavisi ve Uzun Süreli Takibi

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ABSTRACT A 10-year-old female with the chief complaint of severe mobility of the teeth with the red and swollen gingiva was referred to Istanbul University, Faculty of Dentistry, Department of Pedodontics. The patient presented the characteristics of Dentin Dysplasia (DD) I and II. Here is a case report of dentin dysplasia in all teeth, with amorphous type crown and no root or short and malformed roots, associated with severe mobility. Dysplasia type III could be the new entity. The aim of this paper is to present the clinical management of DD and to discuss another type of designation. All teeth were extracted due to hipermobility. Masticatory and esthetic rehabilitation of the patient was performed with total removable dentures. The patient is under regular control since 4 years. Early diagnosis of the condition will help to prevent or delay loss of dentition.

Key Words: Dentin dysplasia; therapy

ÖZET 10 yaşındaki kız hasta, dişlerindeki şiddetli mobilite ve kırmızı, şiş dişeti şikayetleri ile İstanbul Üniversitesi Diş Hekimliği Fakültesi Pedodonti Anabilim Dalı Kliniği'ne yönlendirilmiştir. Olgu dentin displasizi (DD) I ve II'nin karakteristik özelliklerini taşımaktadır. Bu olguda, şiddetli mobilite ile birlikte; amorf kuron, köksüz dişler ya da kısa ve malforme kökler izlenmiş ve tüm dişlerde dentin displasizi olduğu saptanmıştır. Rapor edilen olguda, DD'nin her iki sınıflamasının karakteristik özellikleri birarada görüldüğü için yeni bir sınıflamadan (Dentin displazisi tip III) bahsedilebilinir. Bu makalenin amacı, dentin displazisi saptanan kişilerde tedavi yaklaşımının nasıl olması gerektiğinin ve farklı bir sınıflamanın tartışılmasıdır. Bu olgunun tedavisinde, hipermobiliteye bağlı olarak hastanın tüm dişleri çekilmiştir. Hastanın çiğneme ve estetik rehabilitasyonu hareketli total protez ile sağlanmıştır. Olgu 4 yıldır düzenli olarak takip edilmektedir. Bu durumun erken teşhisi dişlerin erken kaybını geciktirecek veya önleyecektir.

Anahtar Kelimeler: Dentin displazisi; tedavi

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Dentin dysplasia (DD) is a hereditary disorder of dentin formation, marked by a normal appearance of coronal dentin associated with pulpal obliteration, faulty root formation, and a tendency for peripheral alveolar bone lesions without obvious cause. It affects approximately 1:100.000 people.[1-3] Shields et al.[4] sub-classified the condition into two groups; dentin dysplasia, type I and type II. Both are inherited in autosomal dominant manner.[1] Dentin dysplasia type I (DDI) (also known as rootless teeth) primarily affects the root portion of both the deciduous and permanent dentitions.[5-7] DDI, radiographically has short-rooted teeth, typically with sharp conical apical constrictions and crescent-shaped pulpal

remnant parallel to the cemento-enamel junction in permanent dentition and total pulpal obliteration in primary dentition.[8,9] Type II dentinal dysplasia (coronal dentine dysplasia) (DDII) consists of primary teeth with a brown or blue opalascent colour. Radiographically the permanent teeth have large thistle-shaped pulp chambers containing multiple pulp stones.[1,4,8] Ciola et al.,[10] proposed an extension of this classification to include a third group of dentinal dysplasia. Dentinal dysplasia type III (DDIII) includes a combination of teeth of type I and II in the same patient.

O'Carrol et al.[8] categorized DDI, as having 4 radiographic variations in permanent teeth. It was classified into 4 sub-types; 1a, 1b, 1c and 1d. In type 1a, there is a complete obliteration of the pulp and usually little or no root development. Type 1b variation has a horizontal, crescent shaped, radiolucent line, which seperates normal coronal dentine from abnormal radicular dentine. The roots are short, conical and rudimentary. Teeth affected by type 1c variation shows two crescent-shaped horizontal radiolucent lines with their concavities toward each other at the cemento-enamel junction and the roots one half the normal length. Type 1d is characterized by normal root formation, which sometimes may be bulbous in the coronal third. Within the pulpal canal "a stone" may be found. In this type of DD, the pulp chamber is usually not obliterated and normal root formation occurs. This is the least severe form of DD.

Since there is no consensus in the literature regarding the etiology of dentin dysplasia and considering that this is a rare defect of dentin development, case reports can be useful to help elucidating and understanding some aspects related to this condition. This paper reports the case of a child with dental alterations suggestive of both types of DD, in whom most of the teeth showed abnormal occlusal morphology, different features from those found in the literature were observed.

CASE REPORT

A ten-year-old female with the chief complaint of severe mobility of the teeth resulting in discomfort in mastication and aesthetic problems was referred to the Pedodontics Department of Dental Faculty of Istanbul University. Written informed consent had been taken from the parents.

INTRAORAL EXAMINATION

Intraoral examination disclosed presence of the maxillary left first permanent molar, left primary canine, left primary lateral incisor, right primary canine, right primary first and second molars and right first permanent molar (Figure 1). All the upper teeth had caries and excessive mobility. The mandible showed presence of left first permanent molar, primary canine, permanent lateral, central incisors, and right permanent lateral, primary first molar, first permanet molar. Except incisors all the lower teeth had caries. All teeth had excessive mobility. There was no trauma to the teeth. And all teeth present in the maxilla and the mandible except incisors showed abnormal occlusal morphology. Patient's oral hygiene was poor and there were plaque deposits present in all quadrants.

RADIOGRAPHIC EXAMINATION

Radiographic examination (Figure 2a) showed all the teeth in the mandible or maxilla had no root or short and malformed roots. Crown morphology of the molars and premolars were abnormal. In all quadrants first permanet molars and right permanent mandibular second molar had one root. These teeth in the mandible and maxilla appeared to be thistle tube shaped in the coronal pulp and have obliterated pulp in the root canals. 11,13,14,15,17, 21,24,25,27,37,43,47. 24, 27, 37 numbered teeth

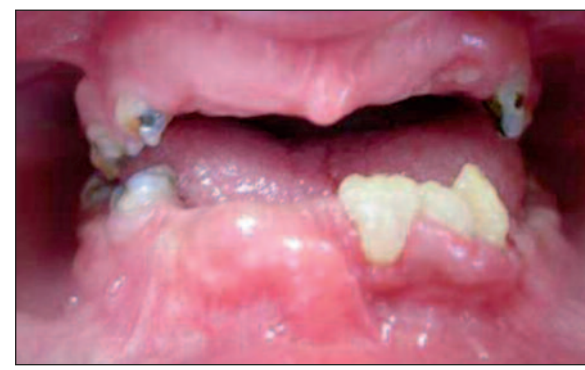

FIGURE 1: Intraoral photograph of the patient.
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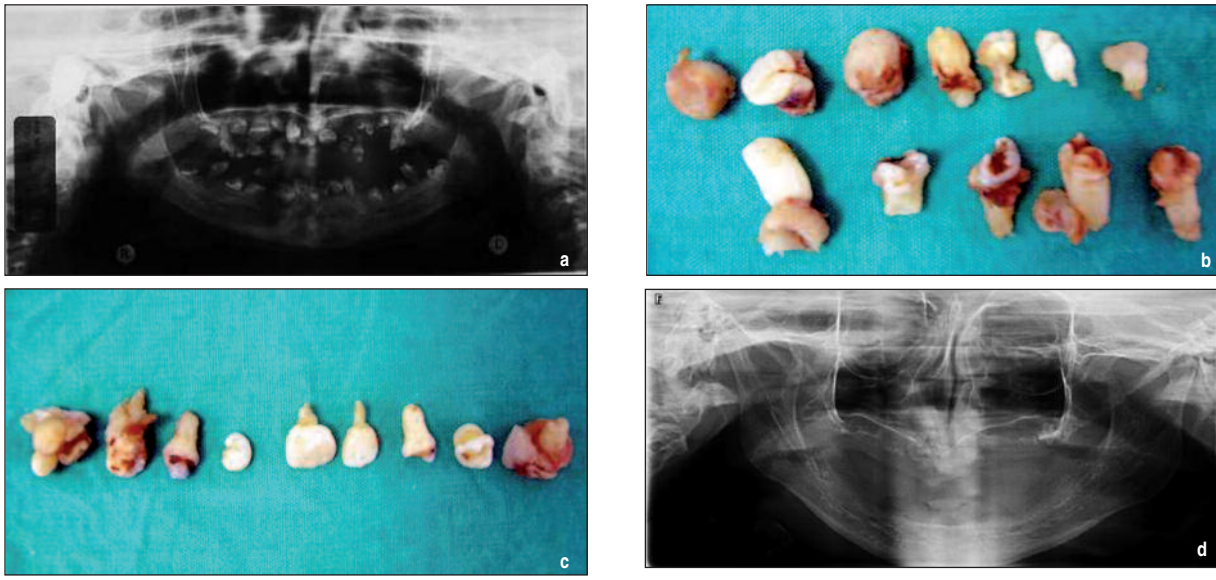

FIGURE 2: a) Panoramic radiograph of the patient before treatment, **b, c)** Photographs of the extracted teeth of the patient **d)** Panoramic radiograph of the patient at 4th year control.
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had hypoplastic enamel with amorphous type crown. Pulp calcification was observed in short-rooted teeth. There was an apical radiolucency associated with 16,26,31,32,36,46. Agenesis of 22,23 and 33 were observed. Primary molars and incisors had periapical inflammation, root resorption was examined. 73 had internal root resorption. Although the patient reached age 10, her maxillary premolars and incisors, mandible premolars and mandible right incisors remained unerupted with no or little root formation. Overextension of the maxillary sinuses was also evident (Figure 2b, c).

HISTOPATHOLOGICAL EXAMINATION

Microscopic examination of longitudinal sections of the extracted teeth were performed by oral pathologists with light and scanning electron microscope. In the decalcified sections regions of hypersementosis, remnants of enamel, dentin and pulp chamber were seen. Occasionally dentin tubuli were regular and there was globular dentin in some areas. The pulp chamber was almost totally occlusied by calcification (Figure 3a, b). Dentin showed atypical tubuler patterns, with amorphous atubuler areas and irregular organization. Dentinal tubules are few in number and they were occlusied by calcification (Figure 3c).

BACKGROUND

She was the sixth child of the consanguineous marriage (Figure 4). According to her mother, the patient was born after an uneventful, full-term pregnancy with no exposure to radiation. The parents also indicated that the teeth of fourth child (female) who died as menengitis at 5-year-old as weak and look like nail. And the presented case was the sixth child of the parents. Birth weight was 3.800 gr. Primary incisors erupted on time but they exfoliated prematurely. Delayed eruption at age 3 and premature exfoliation of the primary molars were observed.

Her mental and physical development was normal according to her age. The extraoral examination showed an abnormal facial appearence, with a dished profile resulting from the loss of numerous teeth. Scars on the face were examined. Endocronologic evaluation for growth hormone, throxin and cortisol were within normal limits. She had no history of any other childhood illness or systemic abnormality.

TREATMENT

Treatment strategy consisted of instructing the patient in oral hygiene, to restore occlusion, enhance

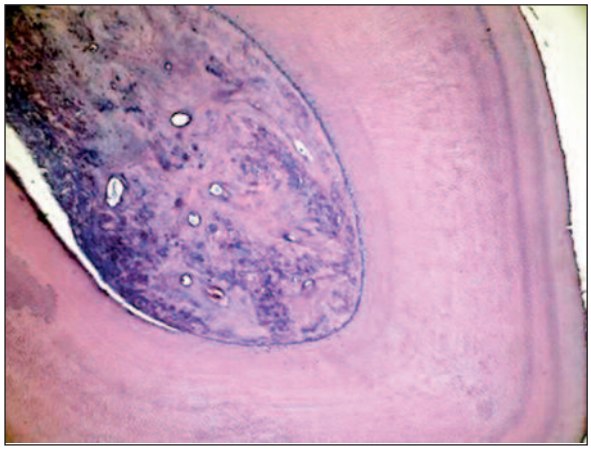

FIGURE 3a: Histopathological examination of the teeth.
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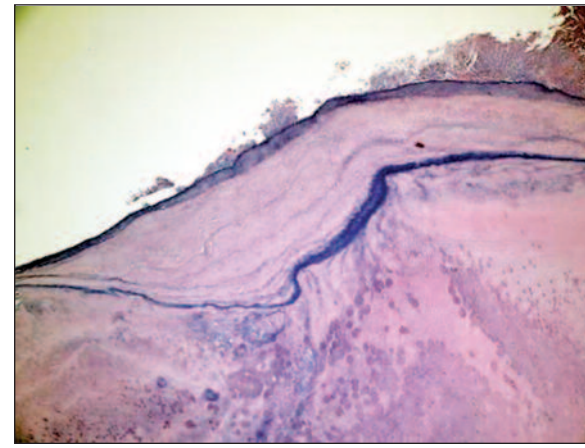

FIGURE 3b: Histopathological examination of the teeth.
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mastication, and improve esthetics. As all the teeth had excessive mobility and periapical inflammation, they were extracted by an oral surgeon (Figure 2b, c). And also unerupted teeth with amorphous crowns, hypoplastic enamel and no root development were extracted under local anesthesia. In order to maintain the proprioception mechanism of the bone, to provide mastication and esthetics, conventional removable dentures were fabricated by a prosthodontist. The patient's main expectation was to improve the reduced chewing ability which was caused by tooth lost. The patient evaluated her function, esthetic and speaking before and 1 year after treatment with VAS score (0-100) (Table 1). Patient's family declared that before treatment patient refused to go to school because of her friends teasing. The dentures improved her facial appearance and jaw functions (Figure 5a, b). The patient was kept under regular control for 4 years. Periodic recall of the young patient was done every 6 months to decide if prosthetic modification or replacement will be needed as a result of continuing growth and development. So, it was decided to change the maxillary and mandibular complete dentures every 6 months serving as interim prosthesis. At 4th year control, radiograhic examination was done (Figure 2d). After radiographic examination, permanent molar tooth on the left side of the maxilla was seen. As the tooth hasn't erupted yet, it was decided to keep under control. The denture of the patient was changed (Figure 5c, d).

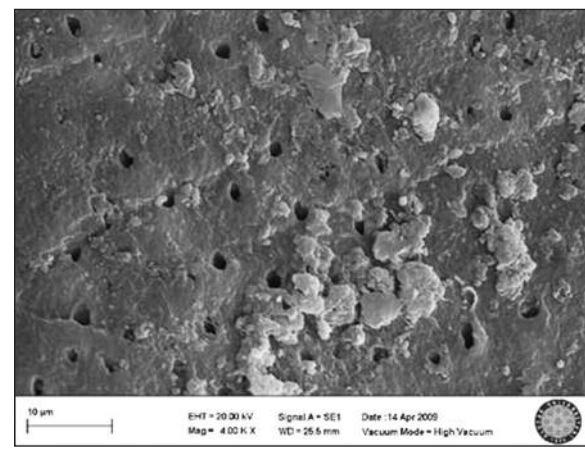

FIGURE 3c: Examination of the teeth with scanning electron microscope.

DISCUSSION

Little is known about etiology of DD in healthy individuals. Several factors have been implicated as possible causes, but the precise nature of the defect has not yet been determined.[3,11] The pathogenesis of dentinal dysplasia has provoked much debate in the dental literature. Logan et al.,[12] proposed that it was the dentinal papillae that was responsible for the abnormalities in root development. They suggested that multiple degenerative foci within the papillae become calcified, leading to reduced growth and final obliteration of the pulp space. Sauk et al.,[13] argued that it is not the dentinal papillae but the epithelial component that is responsible for the calcified foci. They proposed that

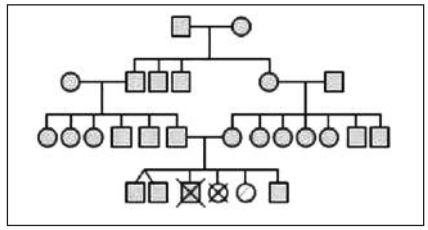

FIGURE 4: Pedigree of the case.

TABLE 1: VAS score (0-100) before and 1 year after treatment.

	Before Treatment (VAS 0-100)	One year after treatment (VAS 0-100)
Function	10	100
Esthetic	0	100
Speaking	30	80

invigilation of the root sheath occurs too early and then makes a sequence of attempts to correct itself. This leaves epithelial foci, which then induces ectopic dentin formation. Wesley et al.,[14] disagree with the theory of Sauks et al. and feel that it is the interaction of odontoblasts with the ameloblast layer that is at fault, and this results in the abnormal differentiation and/or function of these odontoblasts.

In the histopathological examination of the teeth, it was revealed that pulp calcification occurred and irregular dentin was examined. Few number of dentinal tubules were observed in SEM examination. This can be attributed both with abnormalities of development and occlusion of the tubules with distrophic calcification in pulp chamber.[15,16] In the presented case similar histopathological findings were seen.

Most DDI cases have autosomal dominant inheritance, which means that individuals of the same family may be affected.[5,17] However, reports of cases without signs of a family heritage are found in the literature.[6,15]

The patient's parents and siblings were also examined clinically and radiographically to determine whether hereditary transmission had occurred, but no dental alterations were found in the parents and siblings except the patient's deceased older sister whose teeth show similar findings with the case.

Teeth with radiographic or histologic features of DD occur in a number of disorders such as calcinosis, Ehlers-Danlos syndrome, and the Brachioskeletogenital syndrome. Some association has also been reported between dentine dysplasia and osseous changes in addition to sclerotic bone formation but in the case reported by Toomarian et al. had no signs of other pathologic conditions.[15] In this report, she had no history of any other childhood illness or systemic abnormality.

Without radiographic examination, the reported malformation of the teeth might easily be overlooked, in as much as its clinical presentations rather unremarkable. The early exfoliation of the teeth could be mistakenly attributed to more common causes (e.g., poor oral hygiene, periodontitis).[17,18]

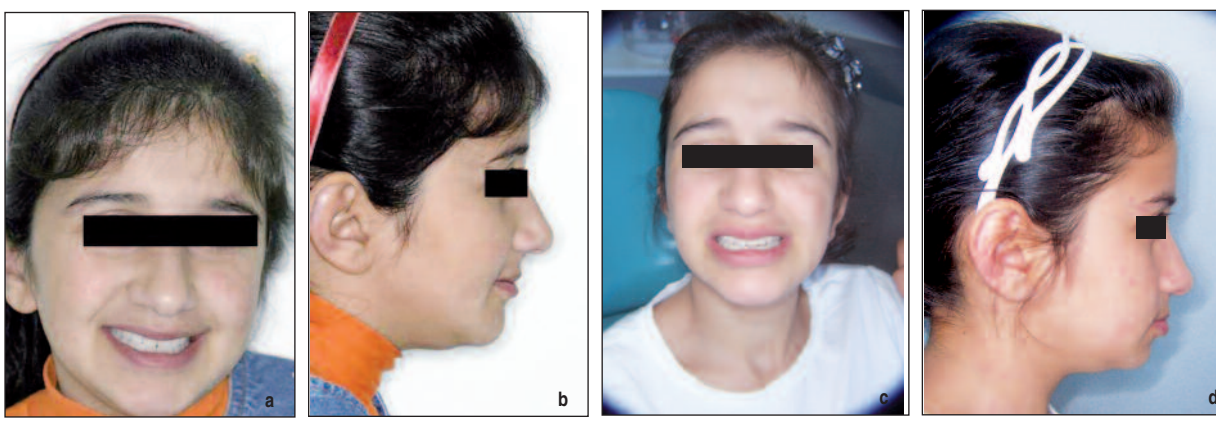

FIGURE 5: a, b) Photographs of the case after first prothetic treatment **c, d)** Photographs of the case after prothetic treatment at 4th year control.
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Although delayed dental eruption has been reported only rarely as characteristic of DDI, it is not suprising to observe a delayed eruption pattern in this disorder, as the development of roots is essential for the eruption of the teeth. Ozer et al.[19] mentioned delayed eruption in two cases with DDI. Kalk et al.[17] observed delayed dental eruption pattern in five cases within one family with DDI. And added that whether a combination of delayed eruption, mobility of the teeth, and incisional opacity could serve as a clinical diagnostic feature in this rare disorder will remain questionable until more cases with similar characteristics have been described.

In this case, the mother of the patient mentioned immediately exfoliated primary incisors and delayed eruption of the primary molar teeth.

In the reported case, rootless teeth or teeth with little root correlate with the sub-classification of DDI type 1a. Radiographic pulp obliteration was observed. Although, in general, teeth affected by DDI have been reported to be of normal shape and size.[6,8] Elzay and Robinson[20] have suggested possible variations in the morphology of the teeth affected by this type of dysplasia. In this case, oppositely as mentioned in DDI, the crowns of the most of the teeth with DDI type 1a, were amorf and hypoplastic. Desai et al reported generalized short-root formation, accompanied by taurodontia, multiple dens invaginatus, microcephalic dwarfism, as well as a prominent feature of dentin dysplasia type I.[21]

The second type of DD (DDII) is characterized radiographically by teeth of nearly normal length but as the pulp ascends to the crown, it flares into a flame shape or thistle tube shape, pulp stones and sudden constriction of the chamber, which forms a thin radiolucent radicular structure.[11,17,18]

Four distinct forms of dentin dysplasia type I and one form of dentin dysplasia type II are identified. Although there seems to be no need to identify more than 2 distinct types of this relatively rare inherited defect of human dentin, the possible existence of additional forms of the disease cannot be ruled out. Dentin dysplasia type III could be the new entity. In all quadrants first permanent molars and right mandibular second permanent molar had one root. The root and pulp of the teeth exposed similar findings with DDI. Opposingly, the crowns of the teeth were hypoplastic and malformed. The findings of the case correlated with both the DDI and DDII, so it could be classified as DDIII.

Recently, a case of dentin dysplasia involving a single tooth was reported. The authors described as focal odontoblastic dysplasia and suggested that reports of other similar cases are probably needed to determine another type of classification (dentin dysplasia type III).[22]

While many cases of DDI have been described, some clinical aspects and theoretic issues remain uncertain. Detailed documentation of clinical presantations in future cases of DDI is needed in order to establish relevant clinical signs that will facilitate the process of diagnosing the disorder. Delayed eruption, variation in occlusal morphology and hypoplastic crowns, unerupted and undeveloped teeth in the maxilla and mandible highlights the clinical and radiographic variations of DDI.[19]

Little is known about the specific treatment of this disorder. The periapical radiolucencies found with the condition are probably secondary to pulpal necrosis, occuring as a result of dental caries or spontaneously, due to exposure of the pulpal remnants through one of the channels in the defective dentin.[23] Spontaneous exfoliation is probably due to periodontal disease occuring on teeth with unfavourable crown-root ratios. Treatment of those teeth that have undergone pulpal necrosis and periapical abcess formation is usually by extraction. Although conventional endodontic therapy has been reported with short term succes, the treatment was carried out on teeth with relatively long roots of DD II morphology.[24,25] A preventive approach to patient management has been emphasized to avoid the periodontal disease and dental caries is the only way to maintain the dentition for an increased period of time.[23,25] However even teeth with no evidence of caries or periodontal disease are lost at an early age in this condition and it seems little can be done despite all the advances in modern-day dentistry.[15,26,27] In this case, rootless erupted

teeth were extracted because of severe mobility. And the unerupted teeth with no roots were also extracted by a surgeon as they will have the same prognosis. In all quadrants first permanent molars and right mandibular second permanent molar that correlate with DDII had severe mobility, periapical abcess, pulp canal obliteration were extracted. In order to maintain the proprioception mechanism of the bone, to provide mastication and esthetics, conventional removable dentures were fabricated. The dentures improved her facial appearance and jaw functions. The patient was kept under regular control for 4 years. Periodic recall of the young patient was done every 6 months to decide if prosthetic modification or replacement will be needed as a result of continuing growth and development. So, it was decided to change the maxillary and mandibular complete dentures every 6 months serving as interim prosthesis.

CONCLUSION

DD is a rare, inherited abnormality of the dentin that may lead to premature exfoliation of the primary and permanent teeth. Early diagnosis of the condition an initiation of effective preventive strategies may help prevent or delay loss of dentition. In conclusion, treatment of a child with DD requires a multidisciplinary approach and through knowledge of behavioral management of the pediatric patient. In this regard, the pediatric dentist assumes an important role in identifying DD and in guiding parents in the selection of measures to prolong the retention of affected teeth.

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REFERENCES

1. Witkop CJ Jr. Hereditary defects of dentin. Dent Clin North Am 1975;19(1):25-45.
2. Gorlin RJ, Cohen MM, Levin SL. Syndromes of Head and Neck. 4th ed. England: Oxford University Press; 2001. p.425-6.
3. Hart PS, Hart TC. Disorders of human dentin. Cells Tissues Organs 2007;186(1):70-7.
4. Shields ED, Bixler D, el-Kafrawy AM. A proposed classification for heritable human dentine defects with a description of a new entity. Arch Oral Biol 1973;18(4):543-53.
5. Ansari G, Reid JS. Dentinal dysplasia type I: review of the literature and report of a family. ASDC J Dent Child 1997;64(6):429-34.
6. Vieira AR, Modesto A, Cabral MG. Dentinal dysplasia type I: report of an atypical case in the primary dentition. ASDC J Dent Child 1998;65(2): 141-4.
7. Naik VV, Kale AD. Dentin dysplasia:single tooth involvement? Quintessence Int 2009;40(3):183-6.
8. Carroll MK, Duncan WK, Perkins TM. Dentin dysplasia: review of the literature and a proposed subclassification based on radiographic findings. Oral Surg Oral Med Oral Pathol 1991;72(1):119-25.
9. Seow WK, Shusterman S. Spectrum of dentin dysplasia in a family: case report and literature review. Pediatr Dent 1994;16(6):437-42.
10. Ciola B, Bahn SL, Goviea GL. Radiographic manifestations of an unusual combination Types I and Type II dentin dysplasia. Oral Surg Oral Med Oral Pathol 1978;45(2):317-22.
11. Hegde M, Hegde ND. Dentin dysplasia-A case report. Endodontology 2004;16(1):16-8.
12. Logan J, Becks H, Silverman S Jr, Pindborg JJ. Dentinal dysplasia. Oral Surg Oral Med Oral Pathol 1962;15:317-33.
13. Sauk JJ Jr, Lyon HW, Trowbridge HO, Witkop CJ Jr. An electron optic analysis and explanation for the etiology of dentinal dysplasia. Oral Surg Oral Med Oral Pathol 1972;33(5):763-71.
14. Wesley RK, Wysoki GP, Mintz SM, Jackson J. Dentin dysplasia type I. Clinical, morphologic, and genetic studies of a case. Oral Surg Oral Med Oral Pathol 1976;41(4):516-24.
15. Toomarian L, Mashhadiabbas F, Mirkarimi M, Mehrdad L. Dentin dysplasia type I: a case report and review of the literature. J Med Case Reports 2010;4:1.
16. Regezi JA, Sciubba J, Jordan RCK. Oral & Maxillofacial Pathology: Clinical-Pathologic Correlations. 5th ed. Philadelphia: WB Saunders; 2008. p.5.
17. Kalk WW, Batenburg RH, Vissink A. Dentin dysplasia type I: five cases within one family. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998;86(2):175-8.
18. Rocha CT, Nelson-Filho P, Silva LA, Assed S, Queiroz AM. Variation of dentin dysplasia type I: report of atypical findings in the permanent dentition. Braz Dent J 2011;22(1):74-8.
19. Ozer L, Karasu H, Aras K, Tokman B, Ersoy E. Dentin dysplasia type I: report of atypical cases in the permanent and mixed dentitions. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2004;98(1):85-90.
20. Elzay RP, Robinson CT. Dentinal dysplasia, reported of a case. Oral Surg 1967; 23(3):338-42.
21. Desai RS, Vanaki SS, Puranik RS, Rashmi GS, Nidawani P. An unusual combination of idiopathic generalized short-root anomaly associated with microdontia, taurodontia, multiple dens invaginatus, obliterated pulp chambers and infected cyst: a case report. J Oral Pathol Med 2006;35(7):407-9.
22. Naik VV, Kale AD. Dentin dysplasia: Single-tooth involvement? Quintessence Int 2009;40(3):183-6.
23. Steidler NE, Radden BG, Reade PC. Dentinal dysplasia: a clinicopathological study of eight cases and review of the literature. Br J Oral Maxillofac Surg 1984;22(4):274-86.
24. Tidwell E, Cummingham CJ. Dentinal dysplasia: endodontic treatment, with case report. J Endod 1979;5(12):372-6.
25. Ravanshad S, Khayat A. Endodontic therapy on a dentition exhibiting multiple periapical radiolucencies associated with dentinal dysplasia Type 1. Aust Endod J 2006;32(1):40-2.
26. Shankly PE, Mackie IC, Sloan P. Dentinal dysplasia type I: report of a case. Int J Paediatr Dent 1999;9(1):37-42.