

Hereditary Opalescent Dentin: A Report of Two Cases

Siddharth Gupta, BDS, MDS; Rahul R. Bhowate, BDS, MDS; Ashok Bhati, BDS, MDS

Abstract

Aim: The aim of this case report is to present the clinical and radiographic findings of hereditary opalescent dentin to facilitate an early diagnosis.

Background: Hereditary opalescent dentin (or dentinogenesis imperfecta) may manifest itself in three variations: i.e., Shields type I, Shields type II, and Shields type III. Dentinogenesis imperfecta occurs as an autosomal dominant trait with variable expressivity, either in presence with osteogenesis imperfecta or as a separate clinical entity in persons who have none of the features of osteogenesis imperfecta.

Case Descriptions: A seven-year old boy and his mother were both diagnosed with hereditary opalescent dentin. A review of the family dental history revealed that this condition affected not only the child's mother but his maternal grandfather and great grandfather. Both the son and the mother exhibited the same clinical and radiologic features as those reported previously with no evidence of osteogenesis imperfecta.

Summary: Being an autosomal disease, hereditary opalescent dentin runs in the family and can affect both the deciduous and permanent dentitions as a dominant trait.

Clinical Significance: Once a patient is diagnosed with hereditary opalescent dentin, other family members should be evaluated given the condition is hereditary.



Keywords: Dentinogenesis imperfecta, opalescent dentin, osteogenesis imperfecta, permanent dentition, deciduous teeth

Citation: Gupta S, Bhowate RR, Bhati A. Hereditary Opalescent Dentin: A Report of Two Cases. J Contemp Dent Pract [Internet]. 2010 July; 11(4):049-055. Available from: <http://www.thejcdp.com/journal/view/volume11-issue4-gupta>

Introduction

Dentinogenesis imperfecta (DI) or hereditary opalescent dentin follows a pattern of non-sex-linked dominant inheritance. This case report describes the impact of this condition involving three generations.

Case Report

A seven-year-old boy, accompanied by his mother, visited the Department of Oral Diagnosis, Medicine and Radiology at the Sharad Pawar Dental College with a chief complaint of rapid wearing of teeth. The teeth were quite fragile and this patient had a history of teeth being worn off rapidly as soon as they erupted. The patient's mother stated she also had a similar oral condition as did her father and grandfather; however, her

brother, sister, and elder son did not. Based on this dental history, the mother also was examined.

On extraoral examination, the sclera of both the child and mother were found to be bluish in color. Intraoral examination of the child revealed that the deciduous teeth were severely worn, almost to the gingival level. The teeth also had a typical yellowish-brown opalescent hue of exposed dentin. Fractured enamel with enamel pits was evident on the occlusal surface of erupted first permanent

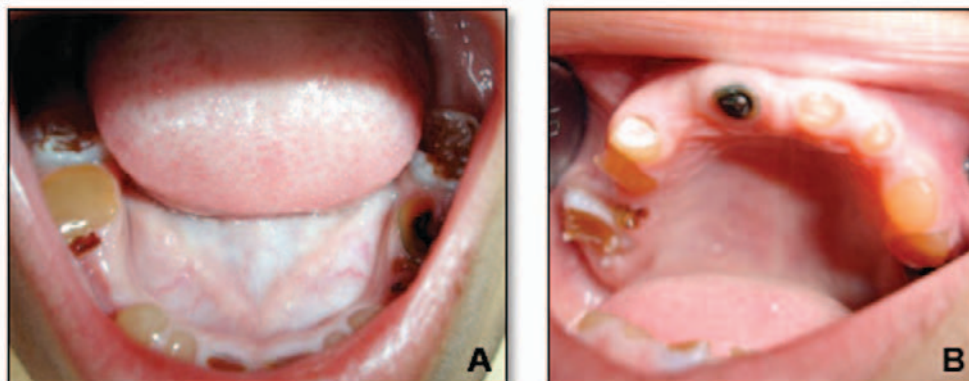


Figure 1. Intraoral presentation of the seven-year-old patient showing evidence of severe attrition of all his erupted teeth.

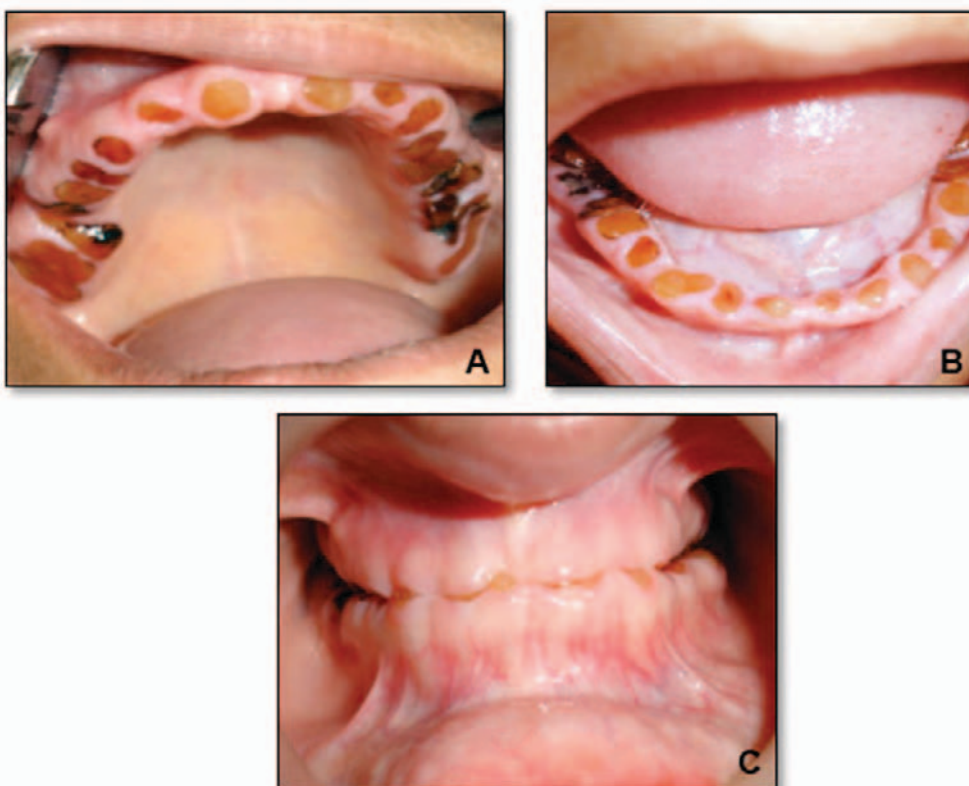


Figure 2. Intraoral presentation of the mother at the initial examination showing severe attrition of her permanent dentition.

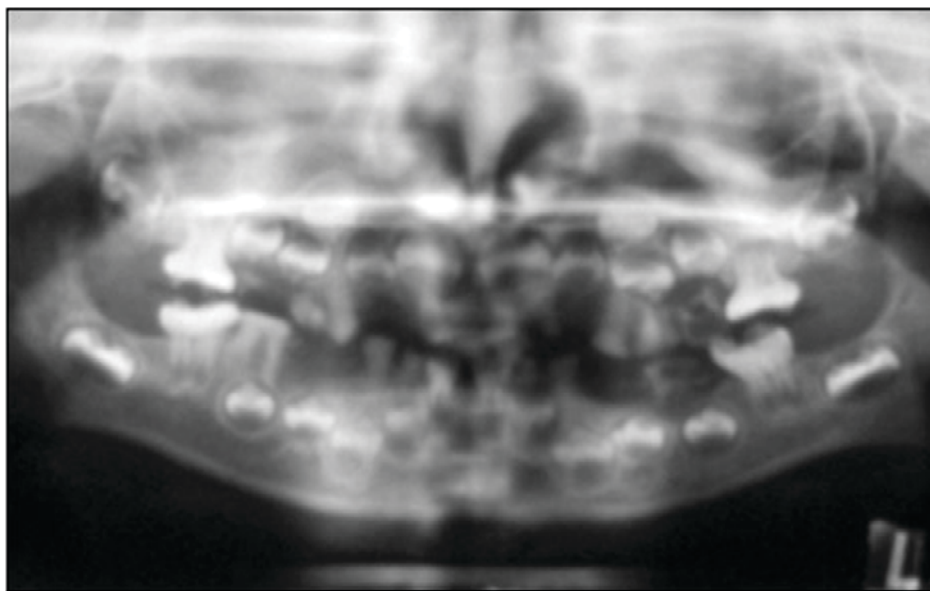


Figure 3. Panoramic radiograph of the seven-year old child.

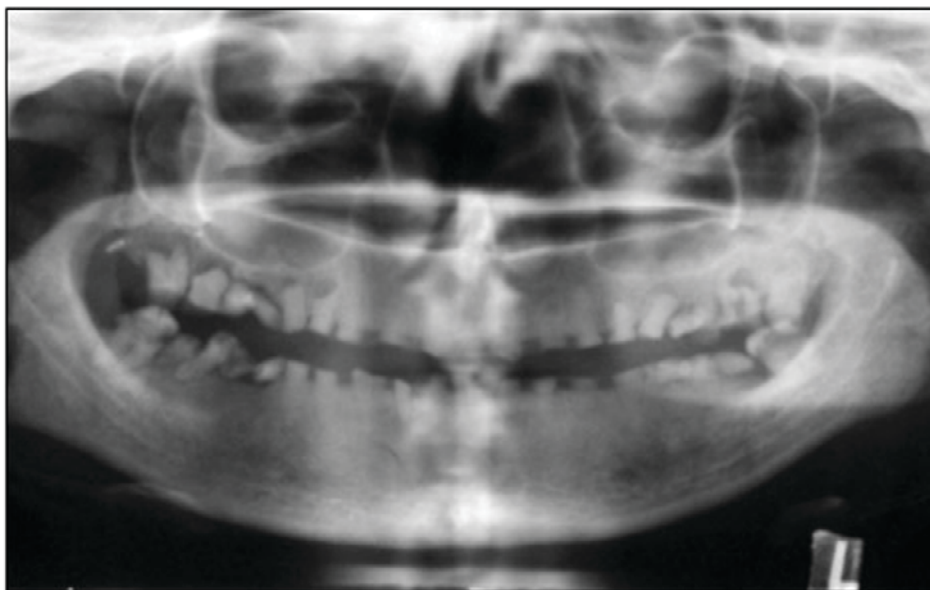


Figure 4. Panoramic radiograph of patient's mother.

molars. The maxillary right deciduous central incisor and mandibular left deciduous second molar were diagnosed with dental caries (Figure 1). Similar levels of attrition and a yellowish-brown opalescent hue to the exposed dentin were seen in the permanent dentition of the mother (Figure 2). Apart from the above-mentioned findings, both patients were in good general health.

A panoramic radiograph of the child showed a mixed dentition with developing permanent teeth (except first molars) and bulbous crown

with cervical constriction of all four permanent first molars. Generalized obliteration of the root canals of the deciduous teeth was seen both in the intraoral periapical radiographs and on the panoramic radiograph. Also, there was decreased radiopacity to all the deciduous teeth (Figure 3). The mother's panoramic radiograph showed a permanent dentition with generalized severe attrition. There was generalized loss of enamel shadow, short blunted roots, and obliteration of the root canal spaces (Figure 4).

Discussion

W.C. Barrett (1882) first recognized opalescent dentin and Talbot (1893) described it as an enamel defect.¹ This condition reportedly occurs in 1 in every 8,000 births,^{2,3} and is caused by a mutation in the dentin sialophospho protein gene (*DSPP*, 4q21.3).^{2,4} The term *dentinogenesis imperfecta* was coined by Robert and Schour in 1939.^{2,5} Witkop and Rao⁶ in 1971 suggested that the term *hereditary opalescent dentin* should be used for isolated traits and the term *dentinogenesis imperfecta* (DI) be used for conditions associated with osteogenesis imperfecta.⁵ Shields et al.⁷ classified the condition into three types:⁶⁻¹⁰

Type I: Dentinogenesis imperfecta in combination with osteogenesis imperfecta (as hereditary mesodermal defect)

Type II: Isolated traits of dentinogenesis imperfecta

Type III: Brandywine isolate of hereditary opalescent dentin⁷

(Type I and Type II are the same, as proposed by Rao and Witkop.⁶)

Clinical features of DI types I, II, and III are almost the same:⁸

- Opalescent color (amber translucency) of both dentitions.
- Bell-shaped clinical crowns.
- Involvement of both dentitions.

- In cases of type III DI, multiple pulp exposures seen in the primary dentition.

Radiographic features of dentinogenesis imperfecta types I and II include obliterated pulp chambers and root canals along with a bulbous clinical crown due to the cervical constriction of the crowns and the short roots.^{6,11} Radiographic features of DI type III may exhibit several variations:⁸

- Normal features, or
- Those features of types I and II dentinogenesis imperfecta, or
- “Shell” teeth, i.e., enormous pulp chamber limited by a thin wall of dentin.^{7,10,12}

The family pedigree chart (Figure 5) for the mother and son actually shows four affected members over four generations. The child’s maternal great grandfather, his maternal grandfather, and his mother were all affected by this condition, but his elder brother and his mother’s elder brother and sister were not. Differentiating features of dentinogenesis imperfecta with amelogenesis imperfecta and dentin dysplasia are explained in Tables 1 and 2.^{5,9}

No characteristic features of osteogenesis imperfecta (i.e., multiple bone fracture, hyperextensible joint, progressive deafness)⁸ and the Brandywine type of dentinogenesis imperfecta (i.e., multiple pulp exposure and shell teeth)⁸ were

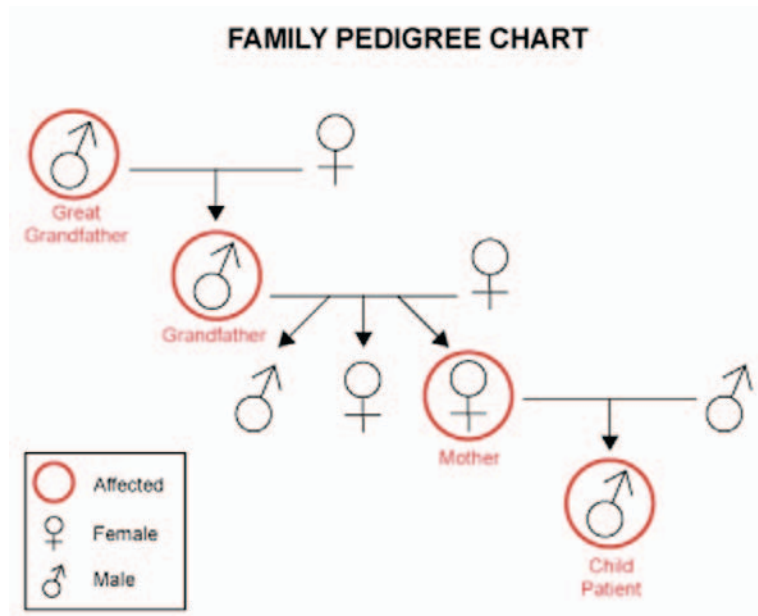


Figure 5. Family pedigree chart showing four affected family members in four generations.

Table 1. Differences between dentinogenesis imperfecta and amelogenesis imperfecta.

Dentinogenesis Imperfecta	Amelogenesis Imperfecta
Affects dentin	Affects enamel
Affects primary and secondary dentitions	Affects primary and secondary dentitions
Normal eruption	Delayed eruption
Fractured enamel with enamel pits	Very thin layer of enamel
Bulbous crown because of cervical constriction of neck	Square-shaped crown
Marked attrition	Less severe attrition
Short and slender roots	Normal roots
Obliteration of pulp chamber	No obliteration of pulp chamber

Table 2. Differences between dentinogenesis imperfecta and dentin dysplasia.

Dentinogenesis Imperfecta	Dentin Dysplasia	
	Shields Type I (radicular)	Shields Type II (coronal)
Comparatively common	Rare	Rare
Opalescent color	Normal to bluish-brown	Primary: amber translucency Permanent: normal
Bulbous crown	Clinically normal teeth with drifting and mobility	Crown shape and size normal
Rarely any periapical lesion	Multiple periapical radiolucencies with noncarious teeth	No periapical radiolucencies
Obliteration of pulp before or after eruption	Obliteration of pulp pre-eruptively in permanent dentition	Obliteration of pulp after eruption with typical thistle tube appearance

observed in the two cases presented. So both cases were diagnosed as hereditary opalescent dentin (dentinogenesis imperfecta type II) with its classic clinical features.

Summary

Being an autosomal disease, hereditary opalescent dentin runs in the family and affects

both the deciduous and permanent dentitions as it is a dominant trait. The child and mother showed many features of hereditary opalescent dentin.

Clinical Significance

Once a patient is diagnosed with hereditary opalescent dentin, other family members should be evaluated given the condition is hereditary.

References

1. Heimler A, Sciubba J, Lieber E, Kamen S. An unusual presentation of opalescent dentin and Brandywine isolate hereditary opalescent dentin in an Ashkenazic Jewish family. *Oral Surg Oral Med Oral Pathol.* 1985; 59(6): 608-15.
2. Kamboj M, Chandra A. Dentinogenesis imperfecta type II: an affected family saga. *J Oral Sci.* 2007; 49(3):241-4.
3. Gage JP, Symons AL, Romaniuk K, Daley TJ. Hereditary opalescent dentine: variation in expression. *ASDC J Dent Child.* 1991; 58(2):134-9.
4. Beattie ML, Kim JW, Gong SC, Murdoch-Kinch CA, Simmer JP, Hu JC. Phenotypic variation in dentinogenesis imperfecta/dentin dysplasia linked to 4q21. *J Dent Res.* 2006; 85(4):329-33.
5. Tanaka T, Murakami T. Radiological features of hereditary opalescent dentin. *Dentomaxillofac Radiol.* 1998; 27(4):251-3.
6. Rao SR, Witkop CJ Jr. Inherited defects in tooth structure. *Birth Defects Orig Artic Ser.* 1971; 7(7):153-84.
7. 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.
8. Johnson R. Odontogenic diseases. In: Lynch MA, Brightman VJ, Greenberg MS, editors. *Burket's oral medicine: diagnosis and treatment.* 8th ed. Philadelphia: J.B. Lippincott Co.; 1984. p. 543-8.
9. Shafer WG, Hine MK, Levy BM. A textbook of oral pathology. 4th ed. Philadelphia: WB Saunders Co.; 2000. p. 58-61.
10. Neville BW, Damm DD, Allen CM, Bouquot JE. *Oral and maxillofacial pathology.* 2nd ed. Philadelphia: Saunders; 2000. p. 94-6.
11. White SC, Pharoah MJ. *Oral radiology: principles and interpretation.* 5th ed. St. Louis: Mosby; 2004. p. 346-7.
12. Clergeau-Guerithault S, Jasmin JR. Dentinogenesis imperfecta type III with enamel and cementum defects. *Oral Surg Oral Med Oral Pathol.* 1985; 59(5):505-10.
13. Mars M, Smith BG. Dentinogenesis imperfecta. An integrated conservative approach to treatment. *Br Dent J.* 1982; 152:15-8.

About the Authors

Siddharth Gupta, BDS, MDS (Corresponding Author)



Dr. Gupta received his master's degree in oral diagnosis, medicine, and radiology from Sharad Pawar Dental College and Hospital of the Datta Meghe Institute of Medical Sciences (Deemed University) in Wardha, India. He is an active member of the Indian Academy of Oral Medicine and Radiology. Presently he is working as senior lecturer in the Career Dental College and Hospital, Lucknow, India.

e-mail: siddharthgupta99@yahoo.com or siddharthgupta4u@indiatimes.com

Rahul R. Bhowate, BDS, MDS



Dr. Bhowate is a professor in the department of Oral Diagnosis, Medicine and Radiology from Sharad Pawar Dental College and Hospital of the Datta Meghe Institute of Medical Sciences (Deemed University) in Wardha, India. He is an active member of the Indian Academy of Oral Medicine and Radiology.

e-mail: dr_bhowate@yahoo.com

Ashok Bhati, BDS, MDS



Dr. Bhati received his master's degree in periodontics from Sharad Pawar Dental College and Hospital of the Datta Meghe Institute of Medical Sciences (Deemed University) in Wardha, India.

e-mail: gums_ashh@yahoo.com