

## Capdepont's Teeth - a Hereditary Dentin Defect. Case Report & Review

**IDientes de Capdepont - un Defecto de Dentina Hereditario.  
Reporte de Caso y Revisión de la Literatura**

**Anuradha Pai<sup>\*</sup>; Shesha Prasad R.<sup>\*\*</sup>; Ramakrishna<sup>\*\*\*</sup> & Raghothama Rao<sup>\*\*\*\*</sup>**

**PAI, A.; PRASAD, R. S.; RAMAKRISHNA & RAO, R.** Capdepont's teeth - a hereditary dentin defect. Case report & review. *Int. J. Odontostomat.*, 6(2):229-234, 2012.

**ABSTRACT:** Dentinogenesis imperfecta is an autosomal dominant genetic disorder with abnormal dentin structure affecting both primary and permanent dentitions leading to discolouration and attrition of teeth. Diagnosis is usually based on family history, a detailed clinical examination and pedigree construction. Treatment involves preservation of teeth, removal of infection, restoration of function and esthetics.

**KEY WORDS:** discolouration, attrition, dentin defect.

### INTRODUCTION

Capdepont's teeth or hereditary opalescent teeth or dentinogenesis imperfecta (DGI) is a hereditary disorder in the absence of any other systemic disorder (Neville et al., 2009) affecting 1 in 8000 births (Bhandari & Pannu, 2008; Barron et al., 2008; Henke et al., 1999; Subramaniam et al., 2008; Yashoda Devi et al., 2011; Kamboj & Chandra, 2007). It shows an autosomal dominant trait affecting both deciduous and permanent dentition (Subramaniam et al.; Shafer et al., 2009; Singh & Singh, 2004). Better understanding and treatment planning of this entity is possible if detected at the primary dentition level.

### CASE REPORT

A 20 year old female patient came to the dental OPD with a complaint of pain in the lower right back teeth since 1 week which was moderate, intermittent, localized, aggravated during night and relieved on medications. She also complained of discoloured teeth since many years.

An abnormal deviation of colour from normal which was evident in her primary teeth, was revealed in history. Medical history for bone fractures, loss of hearing, or any other systemic illness was negative. Family history revealed that her mother had a similar teeth discolouration.



Fig. 1. Severe attrition and grayish discolouration of teeth.

<sup>\*</sup> Professor in Department of Oral Medicine & Radiology, The Oxford Dental College, Bommanahalli, Karnataka, India.

<sup>\*\*</sup> Post-graduate student in Department of Oral Medicine & Radiology, The Oxford Dental College, Bommanahalli, Karnataka, India.

<sup>\*\*\*</sup> Professor in Department of Oral and Maxillofacial Surgery, The Oxford Dental College, Bommanahalli, Karnataka, India.

<sup>\*\*\*\*</sup> Endodontia, Head, RR Multispeciality Dental Care, Karnataka, India.

Intraorally 46, 37, 16, 26 were grossly attrited. 21, 22 showed attrition. Loss of tooth structure till the gingival level was seen in 41, 42, 31, 32, 33. Generalized grayish discoloration was evident (Fig. 1).

Intra oral periapical radiograph in relation to 46 revealed loss of coronal structure involving the pulp with ill defined radiolucency in the periapical region (Fig. 2).

OPG revealed loss of coronal structure with periapical pathology in relation to 16, 26, 31, 32, 33, 41, 42, 37, 46 and 47, partial or complete obliteration of root canals, marked cervical constriction of most of the teeth (Fig. 3).



Fig. 2. IOPAR of 46 revealing periapical infection.

Based on the above findings a diagnosis of dentinogenesis imperfecta type II (Shields classification) was considered. Amelogenesis imperfecta was considered as the differential diagnosis.

Extracted 46 was sent for histopathological analysis which confirmed the diagnosis of dentinogenesis imperfecta (Figs. 4a and 4b).

#### Treatment

Teeth 11 to 15, 21 to 25, 32, 33, 34, 35 and 42, 44 and 45 were endodontically treated and fiber posts were placed and core build up were performed (Figs. 5 and 6). Teeth 16, 26, 37, 31, 41, 46 and 47 were extracted. Fixed partial dentures were planned in upper and lower arches to protect the existing teeth and rehabilitate the missing teeth (Fig. 7).

#### DISCUSSION

A hereditary developmental disturbance of dentin in the absence of any other systemic disorder is termed as dentinogenesis imperfecta (Neville *et al.*). It is a localized mesodermal defect which may affect both primary and permanent dentitions (Bhandari & Pannu; Barron *et al.*; Subramaniam *et al.*; Yashoda Devi *et al.*; Kamboj & Chandra) (as in this case).



Fig. 3. Pre treatment OPG.

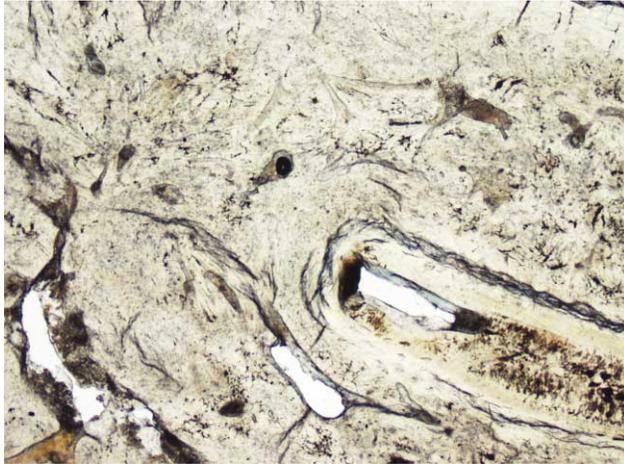


Fig. 4a. Histopathology showing constricted pulp chamber.

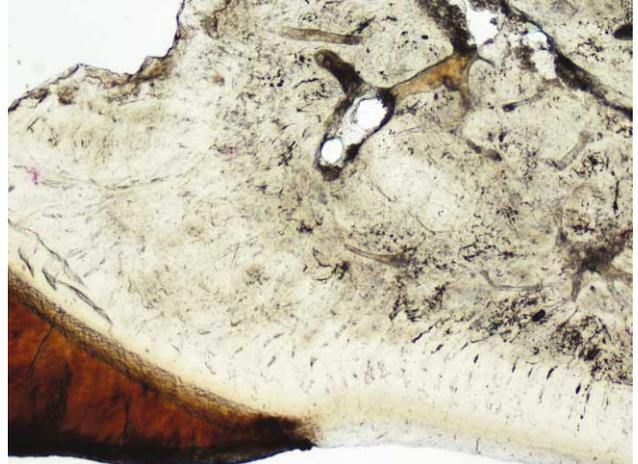


Fig. 4b. Histopathology showing irregular dentinal tubules.

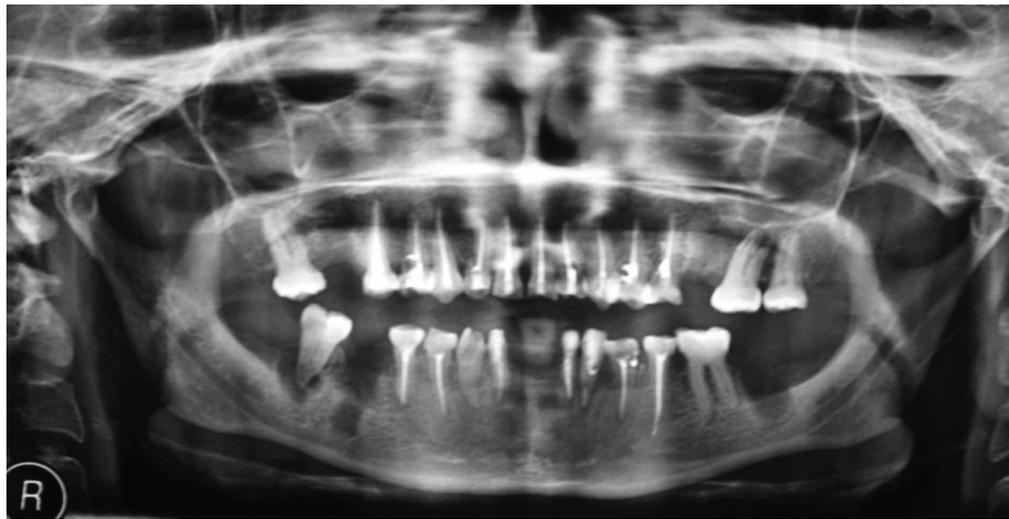


Fig. 5. Post endodontic treatment.



Fig. 6. Core buildup and crown cutting for fixed prosthesis placement.

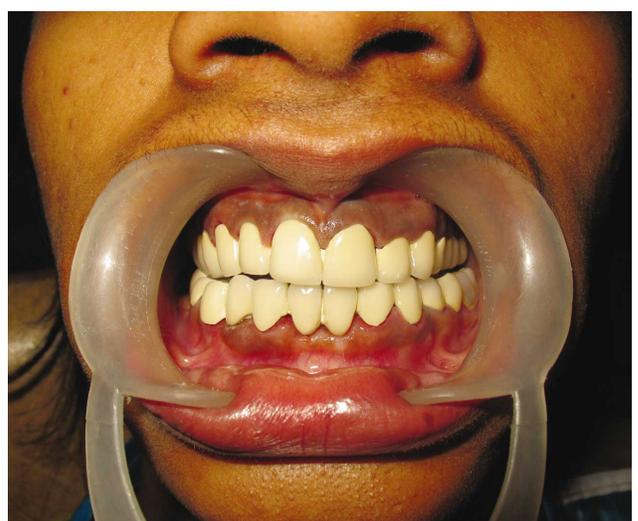


Fig. 7. Post fixed prosthetic restoration.

Barret in 1882 first recognised this condition. Talbot as quoted by Witkop published the first report describing the disorder as an enamel defect. Skillen & Finn used the term hereditary opalescent dentin (Subramaniam *et al.*; Singh & Singh). Roberts & Schour proposed the term dentinogenesis imperfect in 1939. Hursey and associates investigated the racial correlation and reported a wide variation of manifestations of dentin defects (Singh & Singh).

Shields, Bixler and El-Kafraway in 1973 classified dentinogenesis imperfecta into three types.

Type I – DGI associated with osteogenesis imperfect (OI). Both are mesodermal defects, (although OI may occur without DGI).

Type II - DGI without OI.

Type III – Bradywine type. Its a rare variety characterized by shell teeth, with very little dentin and multiple pulp exposures in the primary teeth (Bhandari & Pannu; Barron *et al.*).

The molecular etiologies of the hereditary dentine defects elucidated so far is not accounted by the Shields' system (Barron *et al.*). After years of extensive research it has been proved that DGI is distinct from OI, which called for a revised classification as DGI 1- DGI without OI, corresponding to Shields type II or DGI 2- Bradywine type, corresponding to Shields type III.

However no substitute for Shields type I is provided in this classification (Bhandari & Pannu; Barron *et al.*; Shafer *et al.*).

**Molecular etiology.** Most hereditary dentin defects are secondary to mutations in the genes encoding the major protein constituents of dentin (Barron *et al.*).

DSPP gene (gene map locus 4q21.3) encoding dentin phosphoprotein and dentin sialoprotein (Kamboj & Chandra; Shafer *et al.*), its expression is hundred times higher in the dentin, but is also seen in bone, kidney, salivary gland and lung (Barron *et al.*). Mutations of this gene is the cause for dentinogenesis imperfecta.

From the initially translated polypeptide DSPP three distinct protein products, dentin sialoprotein (DSP), dentin glycoprotein (DGP) and dentin phosphoprotein (DPP) are formed (Barron *et al.*).

Mutations in that region of the gene which encodes DPP results in dentinogenesis imperfecta II, as recently revealed by comprehensive analyses of DSPP. A combination of mis-sense and nonsense changes, splicing mutations and deletions are the overall mutations in DSPP resulting in retention of DSPP as a consequence of errors in signal peptide or subsequent processing events (Barron *et al.*).

DGI I and DGI II are allelic as recent studies state. MacDougall described the intervals separating four genes DSPP, DMP-1, IBSP and SPP1, that are involved in dental development (Shafer *et al.*).

Normal mantle dentin is formed within the dental papilla by the pre odontoblasts that seem to undergo normal cellular differentiation. Gradually due to the mutation in DSPP gene which plays an important role in dentinogenesis these normal odontoblasts are replaced by newly differentiated odontoblasts forming the dental papillae that fail to mature into fully functional odontoblasts secreting an abnormal collagen, which is under mineralized failing to form odontoblastic tubules resulting in dentinogenesis imperfecta (Yashoda Devi *et al.*).

**Clinical features.** Primary teeth are more severely affected than permanent teeth, with incisors and first molars more involved than second and third molars with blue brown, amber or gray discolouration (Barron *et al.*; Subramaniam *et al.*), and distinctive translucency (Neville *et al.*; Bhandari & Pannu). Separation of enamel from defective dentin is noted (Neville *et al.*; Bhandari & Pannu) which when exposed has a glassy, sclerotic appearance (Barron *et al.*), with accelerated attrition of teeth (Neville *et al.*; Bhandari & Pannu; Subramaniam *et al.*), with broader crowns with cervical constriction resulting in tulip shape (Singh & Singh) which were evident in this case.

Over closure of the jaw is a common consequence of tooth attrition seen in dentinogenesis imperfecta which may lead to altered inner ear shape and concomitant hearing deficits as suggested by Kim & Simmer. However the exact cause of the hearing loss cannot be explained (Barron *et al.*). Patient had no hearing defect in this case, also the cardinal signs of flaccid ligaments or fragile bones with previous history of sustained fractures pointing towards associated osteogenesis imperfecta (Singh & Singh) were absent. Hence this case could be categorized as DGI 1 (shields type II)

**Histological features.** The peculiar shade of enamel though normal is the manifestation of the defective dentin with large areas of uncalcified matrix, composed of irregular dentinal tubules, larger in diameter. Readily degenerating odontoblasts which gets entrapped in the dentin matrix is common (Subramaniam *et al.*; Shafer *et al.*; Singh & Singh).

Barring a few subtle hypo calcification defects in the enamel rods just above the DEJ, the enamel appears normal. Although the DEJ appears qualitatively normal it appears flattened (Yashoda Devi *et al.*).

The histologic structure of the mantle dentin appears relatively normal histologically and the characteristic scalloping at the dentinoenamel junction is decreased or missing (Henke *et al.*).

**Chemical and physical features.** Water content is 60% above normal, with less inorganic content than that of normal dentin. The micro hardness of dentin is low and approximates that of cementum, explaining the rapid attrition rate in affected teeth (Bhandari & Pannu; Shafer *et al.*).

**Radiologic features.** Teeth exhibit bulbous crowns with cervical constriction, thin roots and early obliteration of pulp canals (Neville *et al.*; Subramaniam *et al.*; Yashoda Devi *et al.*; Singh & Singh). Some teeth may show normal or enlarged pulp chambers giving it a shell teeth appearance commonly seen in deciduous teeth (Neville *et al.*; Subramaniam *et al.*). Numerous periapical radiolucencies in non-carious teeth (Barron *et al.*) may be seen as in this case.

**Differential diagnosis.** DGI should be differentiated from amelogenesis imperfecta (AI), fluorosis & dentin dysplasia. AI can be recognized by its more clinically apparent enamel defect. The characteristic opalescence or grayish hue is absent. The teeth are usually sensitive with enamel being less radio-dense than dentin on radiographs. Pulp chamber and root canals are usually not sclerosed (Barron *et al.*).

Fluorosis is common in areas of increased fluoride content in water (fluoride belt) with patterns of discolouration that are bilateral, diffuse (not sharply demarcated), opaque, and white striations that run horizontally across the enamel (Abanto Alvarez *et al.*).

Both AI and fluorosis are associated with normal pulp chambers. Normal colouration of both the primary and permanent dentition is present in Dentin

dysplasia type I, although their pulps are almost completely obliterated with extremely short roots and periapical radiolucencies (Yashoda Devi *et al.* & Shafer *et al.*).

Type- II dentin dysplasia does cause a discolored opalescent primary dentition with obliterated pulp chambers. But the permanent dentition is normal in colour and the pulp chambers are enlarged (Yashoda Devi *et al.* & Shafer *et al.*) giving a thistle tube appearance radiologically (Shafer *et al.*).

In congenital erythropoietic porphyria the discolouration ranges from yellow through to green, brown and grey to black and is usually found at the necks of teeth with the enamel hypoplasias usually located in the coronal third of the teeth (Barron *et al.*).

Tetracycline can result in discolouration of both the primary and permanent dentitions. The discolouration varies from yellow or grey to brown and is dose dependent. It also depends on the time of drug intake which stains the teeth according to its rate of development. Alkaptonuria and congenital hyperbilirubinaemia can also cause intrinsic tooth staining (Watts & Addy, 2001) which have other associated signs and symptoms that are absent in DGI. Vitamin D-dependent rickets and vitamin D-resistant rickets can clinically and radiographically mimic DGI (Barron *et al.*).

DGI-I can be a variable feature of Ehlers Danlos syndrome (Watts & Addy), Goldblatt syndrome, Schimke immuno-osseous dysplasia, Brachioskeletogenital syndrome, and osteodysplastic and primordial short stature with severe microdontia, opalescent teeth, and rootless molars (Barron *et al.*).

**Management.** Genetic counseling to parents are important as DGI is inherited as an autosomal dominant pattern. There is a 50% chance that a child born to an affected parent will be affected themselves (Barron *et al.*).

Since the primary dentition is more severely affected in DGI, early diagnosis and treatment is important. Attrition with pulpal involvement, abscesses, rapid decrease in vertical dimension (Barron *et al.*; Sapir & Shapira, 2001) are the problems to be addressed.

Various restorative materials like glass ionomer, composite, polycarboxylate and stainless steel crowns

are advised based on the severity of DGI (Sapir & Shapira). A multidisciplinary approach in collaboration with pedodontist, prosthodontist, orthodontist and endodontist is often imperative in treating mixed and permanent dentition making it challenging (Sapir & Shapira). In this

case, care was taken to retain most of her teeth with endodontic treatment which was challenging owing to the calcified pulp chambers, to ensure proper function. Being young the patient was particular regarding esthetics, which was suitably restored.

---

PAI, A.; PRASAD, R. S.; RAMAKRISHNA & RAO, R. Dientes de Capdepont, - un defecto de dentina hereditario. Reporte de caso y revisión de la literatura. *Int. J. Odontostomat.*, 6(2):229-234, 2012.

**RESUMEN:** La dentinogénesis imperfecta es un trastorno genético autosómico dominante, caracterizado por una estructura anormal de la dentina, que afecta tanto la dentición temporal como permanente, generando decoloración y desgaste de los dientes. El diagnóstico generalmente se basa en la historia familiar, el examen clínico detallado y la construcción de pedigrí. Su tratamiento implica la conservación de los dientes, eliminación de infección, y la restauración de la función y la estética.

**PALABRAS CLAVE:** decoloración, atrición, defecto de la dentina.

---

## REFERENCES

- Abanto Alvarez, J.; Rezende, K. M.; Marocho, S. M.; Alves, F. B.; Celiberti, P. & Ciamponi, A. L. Dental fluorosis: exposure, prevention and management. *Med. Oral Patol. Oral Cir. Bucal*, 14(2):E103-7, 2009.
- Barron, M. J.; McDonnell, S. T.; Mackie, I. & Dixon, M. J. Hereditary dentine disorders: dentinogenesis imperfecta and dentine dysplasia. *Orphanet J. Rare Dis.*, 3:31, 2008.
- Bhandari, S. & Pannu, K. Dentinogenesis imperfecta: a review and case report of a family over four generations. *Indian J. Dent. Res.*, 19(4):357-61, 2008.
- Henke, D. A.; Fridrich, T. A. & Aquilino, S. A. Occlusal rehabilitation of a patient with dentinogenesis imperfecta: a clinical report. *J. Prosthet. Dent.*, 81(5):503-6, 1999.
- Kamboj, M. & Chandra, A. Dentinogenesis imperfecta type II: an affected family saga. *J. Oral Sci.*, 49(3):241-4, 2007.
- Neville, B. W.; Damm, D. D.; Allen, C. M. & Bouquot, J. E. *Oral and maxillofacial pathology*. 3<sup>rd</sup> ed. St Louis, Saunders, 2009. pp.106-8.
- Sapir, S. & Shapira, J. Dentinogenesis imperfecta: an early treatment strategy. *Pediatr. Dent.*, 23(3):232-7, 2001.
- Shafer, W. G.; Hine, M. G. & Levy, B. M. *Shafer Textbook Of Oral Pathology*. 6<sup>th</sup> ed. New Delhi, Saunders, 2009. pp.54-7.
- Singh, M. & Singh, S. Hereditary opalescent dentin--a case report. *J. Indian Soc. Pedod. Prev. Dent.*, 22(3):144-7, 2004.
- Subramaniam, P.; Mathew, S. & Sugnani, S. N. Dentinogenesis imperfecta: a case report. *J. Indian Soc. Pedod. Prev. Dent.*, 26(2):85-7, 2008.
- Watts, A. & Addy, M. Tooth discolouration and staining: a review of the literature. *Br. Dent. J.*, 190(6):309-16, 2001.
- Yashoda Devi, B. K.; Rakesh, N.; Santana, N. & Devaraju, D. Capdepon's Teeth - A Case Report. *J. Indian Dent. Assoc.*, 5(6):753-55, 2011.

Correspondence to:  
Dr. Shesha Prasad  
The Oxford Dental College  
Bommanahalli, Hosur road  
Bangalore 560068  
Karnataka  
INDIA

Email: drsheshaprasad@gmail.com

Received: 16-04-2012

Accepted: 11-07-2012