An integrated treatment approach: A case report for dentinogenesis imperfecta type II

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Abstract
Dentinogenesis imperfecta type II or hereditary opalescent dentin is one of the most common autosomal dominant anomaly of dentin that occurs in both sex affecting approximately 1:8000 persons. Clinically this disorder is characterized by variable blue gray to yellow brown teeth, with fracture of enamel and excessive wear. The treatment strategy is focused towards protecting teeth from further wear and tear and total oral rehabilitation of patient with paramount importance to aesthetics, obtaining an appropriate vertical dimension and providing soft tissue support which will help to return the facial profile to a more normal appearance. A multidisciplinary treatment planning is required for treatment of these individuals.

Key words: Dentinogenesis imperfecta, Opalescent dentin, Oral rehabilitation

Dentinogenesis imperfecta (DI) is a type of heritable abnormality of the dentin that is most common and complicated to manage. Dentinogenesis imperfecta is a genetic disease characterized by a disturbance of dentin formation¹². The primary anomalies of DI are in structure and composition of the dental and result from abnormalities of dentin papilla which is the mesenchymal portion of the tooth germ ³. Clinically these patients have opalescent, amber like teeth which darkens with age and exhibit pronounced attrition of incisal and occlusal edges²⁴. Teeth of both dentition are affected. The other synonyms are Capdepont’s teeth, Hereditary opalescent dentin, hereditary brown teeth etc⁵. Radiographically DI teeth have short roots constricted in their cervical regions and obliterated pulp chambers2,3. Shield’s et al classified dentinogenesis imperfecta based on phenotypic variability⁶⁷.

Type I-DI that occurs with osteogenesis imperfecta. Type II-DI not associated with osteogenesis imperfecta; also known as hereditary opalescent dentin.
Type III-DI of the “Brandywine type”.

Histologically the dentin is irregular with reduced characteristic dentinal tubules. The characteristic scalloping at the dentinoenamel junction is decreased or missing, which is thought to mechanically lock the enamel and dentin and this is the reason attributed for the loss of enamel from the tooth surface ⁸⁻¹¹. Treatment of DI type II requires a multidisciplinary approach with emphasis on occlusal rehabilitation. Aesthetic is an important consideration especially in young patients. Restorative treatment focuses on retaining the remaining tooth structure and protecting the affected dentin from caries, attrition, abrasion and erosion.

Diagnosis and treatment
A 23 years old female came to the Conservative and endodontic department complaining of dull pain in the maxillary anterior region and poor aesthetics due to rapid wearing of the tooth surface. Patient’s medical history was non contributory. The family history suggested that the patients maternal grand father and mother were suffering from the similar condition of rapid wearing of teeth and had undergone total extraction at an young age.

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The clinical examination revealed extensive tooth loss of all anterior and posterior teeth. The remaining tooth structure was brownish in colour. The maxillary anterior incisor teeth were tender to percussion and with associated labial swelling. A detailed history revealed that root canal treatment had previously been attempted on these teeth and not completed. Patient was aware that she had a genetic abnormality and had sought treatment for the same. She was advised to undergo total extraction and have dental implants. Patient did not consider it as a viable option due to which she discontinued the treatment.

Radiographic examination which included full mouth IOPA and OPG showed features characteristic of DI. Roots were short and crown bulbous. The findings of clinical radiographic examination and of family history were consistent with diagnosis of DI. Osteogenesis imperfecta was excluded from the differential diagnosis since there was no associated medical signs and symptoms.

After clinical examination and diagnosis it was recommended that the patient should undergo a full mouth rehabilitation. A treatment plan involving endodontic, prosthetic and aesthetic department was charted out. The initial phase consisted of endodontic therapy for relief of pain and associated symptoms. Complete rehabilitation was planned and the patient was informed.

Radiographic examination of maxillary central incisors revealed periapical abscess in relation to 21. IOPA of the maxillary anterior teeth also revealed that endodontic treatment was attempted with access opening on the central and lateral incisors. Root canals of the maxillary incisors were reopened, irrigated and CA (OH)₂ intracanal medicament was placed. Cleaning and shaping and obturation were completed once the patient was asymptomatic. The next stage of treatment was the restorative and prosthetic phase. Occlusal splints were given to increase the vertical dimension of occlusion for a month, so that future planned raise of vertical dimension in fixed partial denture will be conducive. Restorative treatment included build-up of 23,26,16,15 with composite resin. A prefabricated post was placed on 25 and composite build-up done. Custom made metallic post and core was fabricated and cemented on 11, 12 and 21. On a later date tooth preparation was done on 16, 15, 24, 25, 26, 34, 35, 36, 46 to receive metal ceramic crowns and the maxillary anterior tooth preparation was done for ceramic full crowns. After final upper/lower impression, face bow transfer was done and interocclusal records made. The lower anterior teeth were reduced to stump without any endodontic implication because of the obliteration of the root canal and an over denture was given.

![Fig 1: Preoperative](image1.png)

![Fig 2: Preoperative view in occlusion](image2.png)

![Fig 3: Bite plane](image3.png)

![Fig 4: Posterior metal ceramic crowns and anterior custom made post and core](image4.png)
Discussion
The treatment strategy for patients suffering from dentinogenesis imperfecta should be targeted towards protecting the remaining hard tissues, restoration of stomatognathic system\textsuperscript{12}. There are several alternatives for complete reconstruction of mouth in patients with dentinogenesis imperfecta (DI). Prosthodontic treatment planning includes consideration of function, aesthetics and vertical dimension. In cases involving extensive attrition overdentures or implants are an option\textsuperscript{2}. As in all extensive prosthetic and restorative therapy, the treatment goal is focused on preservation of function and aesthetics.

During treatment planning several factors have to be taken into account, such as age of the patient, quality of the existing dental tissues, the periodontal condition, pulp root anomalies, the loss of tooth structure and orthodontic condition\textsuperscript{13}. Depending on various factors the treatment options may vary. The distinctive radiographic appearance of DI is critical in establishing the exact diagnosis. The difficulty or impossibility of performing endodontic treatment due to the anatomy of the DI affected teeth makes treatment planning more critical\textsuperscript{12}.
Endodontic treatment when needed to assist with restorative treatment, should be considered. Despite radiographic obliteration the pulps of DI teeth can remain vital and do not typically necrose because of overproduction of dentin. The increase in risk of calcification in such teeth can justify the need for prophylactic endodontic treatment for strategic teeth. Early elective endodontic treatment is a viable approach that may improve long time prognosis. In this patient endodontic treatment was complicated by the fact that there was a previous unsuccessful attempt to endodontically treat the teeth, leading to an increased chances of perforation because of improperly prepared canals. Endodontic hand instrumentation was used for more tactile feel of the canal to avoid canal perforations. Obturation was completed using conventional lateral condensation technique. Due to extreme attrition there was lack of vertical height and improper occlusion which was corrected before restorative treatment was commenced. Complete coverage crowns are the preferred restoration for these patients, because such restorations protect the dental tissues from further destruction.

Conclusion
Total rehabilitation of the patient with dentinogenesis imperfecta is a challenge to the dental surgeon which requires the active involvement of various branches of dentistry. Oral rehabilitation of this type of condition should be oriented towards functional and aesthetic rehabilitation.

Reference