Dentinogenesis Imperfecta: A Case Report

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ABSTRACT:
Dentinogenesis imperfecta (DGI) is an autosomal dominant disorder which is characterized by defective dentine formation. The defect arises in the histodifferentiation stage during the tooth development. The teeth appear opalescent. The condition may affect the primary or permanent dentitions or both. The condition may or may not be found in association with osteogenesis imperfecta. This condition has a reported incidence of 1/6000 to 1/8000. There are three sub-types namely type I (DGI with osteogenesis imperfecta), type II (DGI without osteogenesis imperfecta) and type III (DGI associated with Brandywine triracial isolates). Treatment involves multidisciplinary approach to provide optimal oral health, restoration of function and aesthetics.

Keywords: Endodontic failure, Retreatment, Variations in dental anatomy.

INTRODUCTION
Dentinogenesis imperfecta is a hereditary disorder of dentin formation resulting in abnormal dentin formation.1 It was first reported by Barret in 1882.2 It is in accordance to autosomal dominant Mandelian trait.3 Synonyms are Hereditary opalescent dentin due to clinical discoloration of teeth or candeont dysplasia.4 Molecular etiology is found to be related with dentine sialo phosphoprotein gene (DSPP). DSPP is located within the chromosome 4q22.1 in human. DSPP expression is hundred times higher in dentin, it is expressed in bone, kidney, salivary glands as well.1 Absence of microscopic scalloping which is normally seen between dentin and enamel is reported. Due to this, the dentin gets easily separated from the enamel resulting in attrition.5 DGI may affect both the dentitions but has preponderance towards the primary dentition. The permanent teeth which erupt early are found to be more affected.8 Clinically, the color of the teeth varies from brown to blue, amber or grey, with an opalescent shine.9 The crowns are bulbous with cervical constriction. Radiographically, Obliteration of pulp chambers and root canals due to secondary dentin deposition are seen. Normal roots or short roots are seen. Sometimes, root may be absent.1,8

CASE REPORT
A 32-year-old female patient reported to the department of oral medicine and radiology with a chief complaint of sensitivity in all her teeth since 2 months. She complains of sensitivity on eating cold, sweet and sour food items. Sensitivity lingers for around 2-3 minutes after removal of the stimulus. History revealed that her primary dentition had a similar yellowish discoloration. Her mandibular right and left back teeth were extracted as the teeth were decayed. Her family and social history was non-contributory. No history of consanguineous parents reported. There was no history of any unusual bone brittleness or any other systemic illness. Extra-oral examination was non-contributory.
On intra-oral examination, all maxillary teeth were present whereas mandibular right and left posterior teeth were missing. Root stump of maxillary left canine was seen. Generalized yellowish discoloration of teeth was seen (Figure 1). Generalized attrition was present. The maxillary left first premolar and second molar were carious. Also, mandibular left premolars and right third molar were found to be decayed. Investigations advised were intraoral periapical radiographs (Figure 2), Orthopantomogram (Figure 3) and cone beam computed tomography scan (Figure 4) for prosthetic rehabilitation.
On the basis of clinical examination, a provisional diagnosis of dentinogenesis imperfecta was made. The intraoral periapical radiographs revealed complete loss of enamel and obliteration of pulp space, bulbous crowns with cervical constriction. Orthopantomogram revealed generalized loss of enamel. Cone beam computed tomography scan was non-contributory in the diagnosis but helped in the evaluation for the implant placement.

The treatment plan advised included prosthetic rehabilitation and restoration of the decayed teeth. Also, extraction of the maxillary left canine (root stump) was advised.

DISCUSSION
Kerbel et al.,10 and Wright et al11 showed abnormal dentinal tubules with defect in the dentinal calcification. Though the dentin structure found to be defective, the enamel and other structures such as cementum, periodontal ligament are normal. Due to defective dentin deposition, leading to defective dentinoenamel junction and hence the easy chipping of the enamel. The transparent appearance of the dentin is due to reduced number of dentinal tubules or complete absence of dentinal tubules. There is less of calcium (Ca), phosphorus (P), magnesium, a higher Ca:P ratio, and higher water content within the affected dentin. The main cause for premature fracture in DGI is the lower mineral concentration as compared to normal dentin.15

Shields and co-workers classified DGI into three types:13
Type I - DGI in association with osteogenesis imperfecta
Type II - DGI without osteogenesis imperfecta
Type III – Brandywine type. It is associated with Brandywine triracial isolates and characterized by shell teeth, with very less dentin and multiple pulp exposures in the primary teeth.14
A revised classification was given on the basis of the extensive research that has shown DGI and osteogenesis imperfecta are two different unrelated entities. The classification is as follows:

- DGI I corresponds to DI type II
- DGI II corresponds to the DI type III of Shields classification, respectively.

In the present case, the absence of osteogenesis imperfecta and any other systemic disease makes the diagnosis of DG I II favorable. According to revised classification, present case falls under the category DGI I. Also, generalized yellowish discoloration, distributed in all the four quadrants was seen. It was observed that generalized attrition of the crowns caused a decrease in vertical dimension.

Treatment includes protection of the teeth from attrition, functional rehabilitation and aesthetic improvement. The treatment varies according to the patient’s age and presenting complaint. If the primary teeth are involved, stainless steel crowns may be used and to improve the aesthetics, composite facing crowns plays a crucial role. Overdentures may be given in conditions when severe attrition is present. If the permanent teeth are involved, preservation or restoration of the vertical dimension is mandatory. Importance should be given to minimal tooth preparation until child reaches adulthood. Full mouth rehabilitation is generally the choice of treatment in case of severely affected teeth. Endodontic treatment may be difficult to perform as obliteration of pulp chamber are seen. Prognosis is questionable even after endodontic treatment due to the increased probability of fracture.

CONCLUSION
Due to the excessive wear and premature micro injuries, the treatment planning is the most important factor. Treatment is multidisciplinary which includes pedodontics, prosthodontics, orthodontics, endodontics. Full mouth rehabilitation is required to improve the condition and health of the supporting structures. Early diagnosis and long term follow up plays an important role to prevent the early teeth wear and to intercept at the earliest.

REFERENCES