Dentinogenesis Imperfecta (Hereditary Opalescent Dentin) in Primary Dentition: A Case Report

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ABSTRACT

Dentinogenesis imperfect (DGI) is an autosomal dominant disorder of tooth development which is characterized by the presence of opalescent dentin, which leads to dusky blue to brownish discoloration of the teeth. Enamel is generally thinner than normal in this condition due to abnormal scalloping at dentino-enamel junction, thus the teeth are also weaker than normal. This makes them more susceptible to rapid attrition, breakage, and loss. In most of the cases, an affected person has atleast one parent with this condition. This article reports a case of dentinogenesis imperfect (DGI) in a 06 year old female with typical clinical and radiographic features and family history.

KEYWORDS: Abnormal Dentino-Enamel Junction, Dentinogenesis Imperfecta, Hereditary Opalescent Dentin

INTRODUCTION

Dentinogenesis imperfect was probably first documented by Barret in the year 1882. The first report describing this disorder as an enamel defect was by Talbot which is cited by Witkop.[1] The term 'hereditary opalescent dentin' was first reported by Skillen,[2] Finn [3] and Hodges[4] to define the brown translucent teeth that have an opalescent sheen and are lacking in pulp chambers. Dentinogenesis imperfecta is a localized mesodermal dysplasia which affects both the primary as well as the permanent dentitions. It is inherited in an autosomal dominant pattern with high penetrance and a low mutation rate.[5] It is one of the most prevalent dental genetic disease; affecting approximately 1 in 8000 births.[6] The teeth may vary in colour from brown to blue, sometimes pronounced as amber or gray. Enamel is generally thinner than normal in this condition due to abnormal scalloping at dentino-enamel junction, thus the teeth are also weaker than normal. This makes them more susceptible to rapid attrition and breakage.

CASE REPORT

A 06 year old girl patient reported to the department of Paedodontics and Preventive Dentistry with the chief complaint of discoloration of teeth. Intraoral examination showed generalized yellowish- brown discoloration (Fig. 1) and generalized attrition of teeth (Fig 2a, 2b). Her family history revealed that, her grandfather, father, paternal uncle and her paternal aunty had similar discoloration of the teeth.

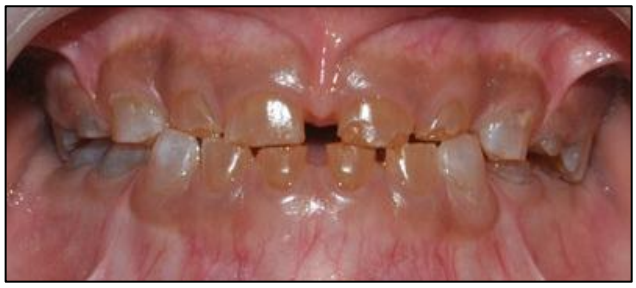

Fig 1 show generalized yellowish-brown discolouration of teeth

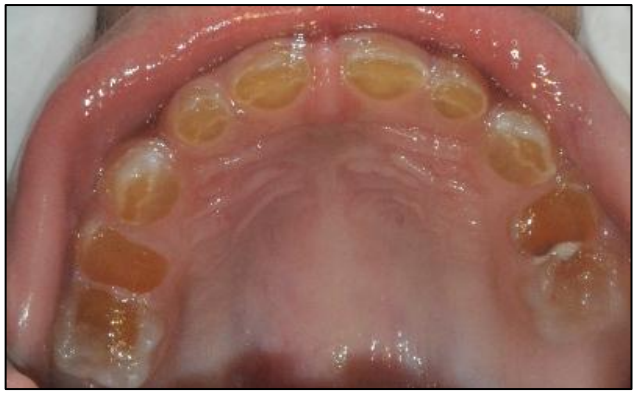

Fig 2a shows generalized attrition in maxillary arch.

How to cite this article:
Shetty RM, Goyal A, Kandelwal M, Deoghare A, Hanumanta S, Parakh K. Dentinogenesis Imperfecta (Hereditary Opalescent Dentin) in Primary Dentition: A Case Report. Int J Dent Med Res 2015;1(5):87-88.

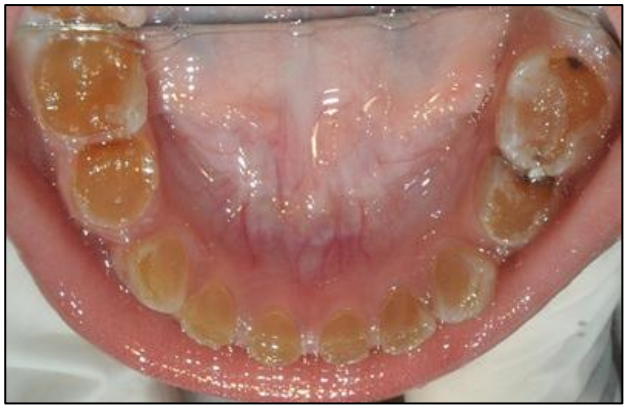

Fig 2b shows generalized attrition in mandibular arch

Orthopantomogram findings revealed that posterior teeth have shorter and slender roots with neck being constricted. Generalized obliteration of pulp canal with multiple pulp stones were also noticed (Fig 3).

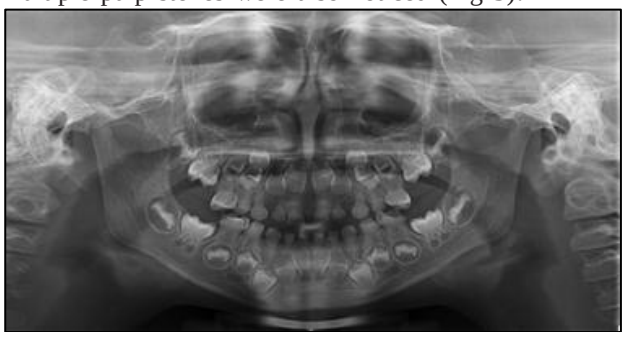

Fig 3 shows short and slender roots, constricted neck, prominent in posterior teeth, increased contrast between crown and root, and generalized obliteration of pulp canal.

Diagnosis of DGI type I was made on the basis of history, clinical examination, radiographic findings, and the autosomal dominant inheritance pattern in the family over three generations.

DISCUSSION

According to Shield[5] Dentinogenesis imperfect is subdivided into three types: type I which is associated with osteogenesis imperfect (OI); in type II where there is no associated osteogenesis imperfect and when the condition is being associated with the Brandywine tri racial isolate and larger pulp chambers it is classified under type III.

Extensive research over the year has proven that DGI and OI are two separate entities, isolated from each other. A revised classification has been suggested where DGI is classified as type I and II. And both types are not associated with OI. DGI I corresponds to DGI type II and DGI II corresponds to the DGI type III of shield classification respectively. There is no substitute for DGI type I in this revised classification.[1] According to revised classification the reported case is affected with DGI type I. The teeth affected with DGI type I emphasize purely a mesodermal defect in which the primary structural abnormality is seen in dentin. Kerebell et al.[6] showed gross abnormality in dentinal tubules and dentinal calcification where as other structure like enamel, cementum and periodontal ligament were normal. The trait shows 100% penetrance but variable expressivity which was also seen in the reported case. According to the family history given by her all the affected family members had varying expressivity of the disorder. In this case there was generalized brownish discoloration attrition whereas her father exhibited a grayish discoloration only in his lower anteriors. Wearing of the enamel is supposed to be due to the decreased mineralization of dentin along with higher water content and abnormal dentinoenamel junction. This was also reported by Kinney *et al.*[7] that DGI type I is an autosomal dominant mutation in dentin sialophosphoprotein gene (Gene map locus 4q12-q21) which encodes for two dentin specific noncollagenous acidic matrix proteins: dentin sialophosphoprotein (DSP) and dentin phosphoprotein (DPP). These genes constitute 50% of the noncollagenous composition of dentin. The affected dentin has decreased calcium, phosphorus, magnesium but increased Ca: P ratio with higher water content. The chief characteristic of DGI type I is higher wear rate due to the absence of intrafibrillar mineralization. Lower mineral concentration in dentin may leads to the premature fracture of teeth.[7] According to Kinney et al.[7] decrease or absence of dentinal tubules are responsible for giving the appearance of opalescent dentin. Opalescent dentin was revealed in the present case too.

CONCLUSION

Early diagnosis and appropriate treatment is of paramount significance to prevent psychological and functional morbidity to the patient. A comprehensive interdisciplinary treatment planning is required to rehabilitate patient affected with DGI. As in this case there is no functional loss, the occlusal was stable patient is kept under regular follow up for any intervention if required.

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Source of Support: Nil
Conflict of Interest: Nil