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CASE REPORT

DENTINOGENESIS IMPERFECTA TYPE II: A CASE REPORT

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ABSTRACT

Dentinogenesis imperfecta or capdepont's teeth is an autosomal-dominant trait with variable expressivity. It affects dentin of both the primary and permanent dentitions. This condition is also called as hereditary opalescent dentin because of clinical discolouration of teeth. In this case report we are presenting a case of dentinogenesis imperfect type II with the disease affecting two generations of a family. The purpose of this report is to highlight the features of this disease and to differentiate it from its other variables. Early diagnosis and intervention with preventive strategies are recommended to prevent the deterioration as well as to improve the dental aesthetics and general health of the patient.

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INTRODUCTION

Dentinogenesis imperfecta (DI) is a disease characterized by an abnormal formation and thus abnormal structure of the dentin, generally affecting both primary and permanent dentition. The prevalence of dentinogenesis imperfecta is 1 in 6,000 to 8,000. (Barron *et al*., 2008) There are three types of DI (Table 1) (Barron *et al.,* 2008).

Dentinogenesis Imperfecta Type I (DI-I)

Characterized by presence of features of osteogenesis imperfecta. The teeth of both dentitions are typically amber coloured, translucent with significant attrition. Radiographically, the teeth have short, constricted roots and dentine hypertrophy leading to pulpal obliteration either before or just after eruption. While some teeth show total pulpal obliteration in others the dentine appears normal (Barron *et al.,* 2008).

Dentinogenesis Imperfecta Type II (DI-II)

The dental features are similar to DI-I but genetic penetrance is virtually complete and osteogenesis imperfecta is absent. Hearing loss has also been reported as a rare feature of this condition (Barron *et al.,* 2008).

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Dentinogenesis Imperfecta Type III (DI-III)

This form is most commonly seen in a tri-racial population from Maryland and Washington DC known as the Brandywine isolate. The clinical features are; the primary teeth show multiple pulp exposures and radiographically, they often manifest as "shell" teeth i.e. hollow teeth due to hypotrophy of the dentine (Rocha *et al.,* 2011). Other features are similar to DI- I and II.

Case report

An 8 year old girl reported to the Department of Paediatric Dentistry with the chief complaint of multiple carious teeth. Her medical and dental history was non-contributory. She had a history of extraction in lower jaw due to extensive carious lesions. Her general examination revealed no abnormality. Intraoral examination revealed translucent amber colour of teeth with slight enamel chip off. (Fig. 1) Based on clinical findings our provisional diagnosis was Dentinogenesis Imperfecta. With the differential diagnosis of Amelogenesis Imperfecta (AI), and Dentin Dysplasia (DD). Panoramic examination revealed presence of bulbous crowns, cervical constriction, pulp obliterations in some teeth and periapical radiolucency; in addition to that both the lower second premolars were congenitally missing. (Fig. 2) Based on the clinical and radiographic features our final diagnosis was DI-II. The treatment plan was formulated as per AAPD 2004 objectives (Sapir *et al.,* 2001).

Table 1. Classification of Dentinogenesis Imperfecta

Shields	Clinical presentation	Witkop
Dentinogenesis imperfecta I	Osteogenesis imperfecta with opalescent teeth	Dentinogenesis imperfecta
Dentinogenesis imperfecta II	Isolated opalescent Teeth	Hereditary opalescent teeth
Dentinogenesis imperfecta III	Isolated opalescent teeth	Brandywine Isolate

Table 2. Comparison of Clinical and Radiographic Features of Dentinogenesis Imperfecta and Dentin Dysplasia

Dental features	Dentinogenesis imperfect I	Dentinogenesis imperfecta II	Dentinogenesis imperfecta III	Dentin dysplasia I	Dentin dysplasia II
Bulbous crowns	-	+	+	-	+
Cervical constrictions	-	+	+	-	+
Periapical radiolucencies	-	-	+	+	-
Short constricted roots	+	+	+	-	+
Thistle tubed pulp chamber	-	-	-	-	+
Primary teeth affected	+	+	+	+	+
Permanent teeth affected	+	+	+	+	-
Amber translucent (opalescent) colour	+	+	+	-	+
Shell teeth	-	-	+	-	-
Normal roots	-	-	-	-	+
Attrition	+	+	+	-	+
Osteogenesis imperfecta	+	-	-	-	-
Rootless teeth	-	-	-	+	-

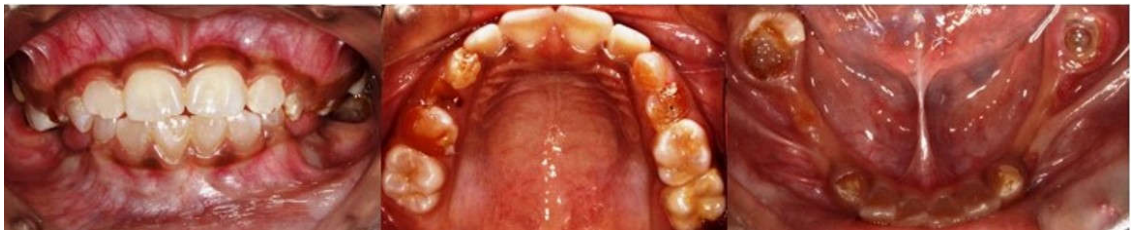

Figure 1. Intra-oral photographs showing translucent amber coloured early mixed dentition

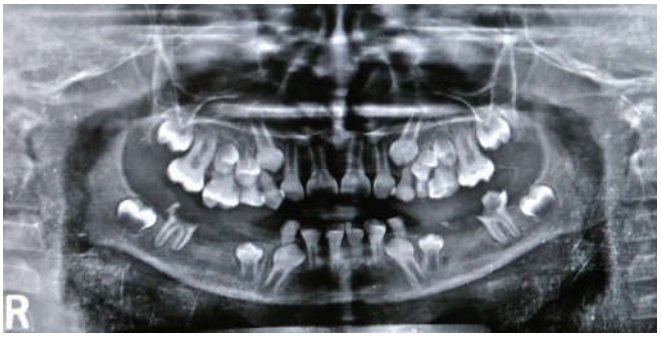

Figure 2. Panoramic radiograph showing bulbous crowns, cervical constriction, pulp obliterations, with minimum pulp remaining and congenitally missing 35, 45

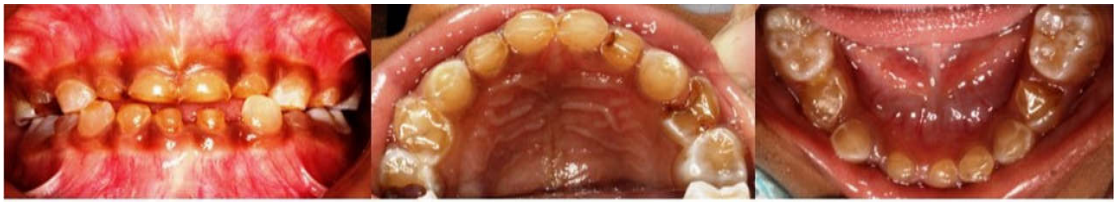

Figure 3. Intra-oral photographs of affected sibling showing translucent amber coloured primary dentition with attrition

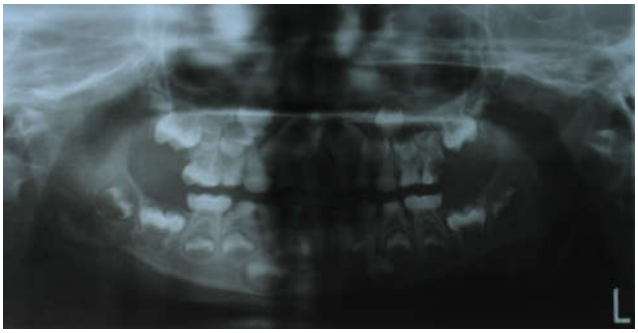

Figure 4. Panoramic radiograph showing bulbous crowns, cervical constriction, pulp obliteration and minimal pulp remaining

Affected Sibling

Family history revealed that similar amber coloured teeth were present in her younger brother's oral cavity. His medical and dental history was non-contributory. Clinical examination revealed generalised attrition with translucent amber colour of teeth. (Fig. 3) His panoramic radiograph revealed presence of bulbous crowns, cervical constriction and pulp obliteration in some teeth. (Fig. 4)

DISCUSSION

Dentinogenesis imperfecta was first recognized by Barret in 1882 (Nayar *et al.,* 1981). The term was coined by Robert and Schour in 1939 (Kamboj and Chandra, 2007). DI type II, III and Dentin Dysplasia type II (DD II) are considered as allelic with variable expressivity because they appear to result from mutations in the same gene that is dentine sialo-phospho-protein (DSPP). The DSPP gene provides instructions for making two proteins that are essential for the formation of dentin (Neville *et al.,* 2007). Clinically, all three types of dentinogenesis imperfecta share numerous features (Table 2) (Neville *et al.,* 2007). As seen in our case both dentitions exhibit an unusual translucent appearance with amber colour. The entire crown appears discoloured because of the abnormal underlying dentin. Although the enamel is structurally and chemically normal, it fractures easily, resulting in rapid wear due to the poor support provided by the abnormal dentin and possibly in part to the absence of the microscopic scalloping normally seen between dentin and enamel that is believed to help mechanically lock the two hard tissues together (Neville *et al.,* 2007). Radiographically opacification of dental pulp occurs because of continued deposition of abnormal dentin.

The short roots and the bell-shaped crowns are also obvious on radiographic examination. Microscopically, the enamel appears to be normal but dentin contains fewer, but larger and irregular, dentinal tubules. The pulpal space is nearly completely replaced over time by the irregular dentin (Regezi *et al.,* 2003). Differential diagnosis includes hypo-calcified forms of amelogenesis imperfecta, which is characterised by poorly calcified enamel which is soft and friable and is rapidly lost by attrition leaving dentine cores. Unlike DI the teeth are usually sensitive and enamel is less radio-dense than dentine. Pulp chamber and root canals are usually not obliterated (Garg *et al.,* 2012). DD-II was ruled out because of the absence of "Thistle Tube appearance" and involvement of Permanent dentition (Neville *et al.,* 2007).

DI may be associated with systemic diseases; Such as Ehlers-Danlos syndrome and Gold blatt syndrome (Singhal *et al.,* 2014). From the point of view of dental care, treatment is directed towards improving the aesthetic appearance with protection of the existing teeth from wear. Generally, fitting with full crowns at an early age is the treatment of choice.

Conclusion

For the diagnosis of DI tooth dis-colouration and syndromes related to it should be ruled out as early diagnosis will help minimise nutritional deficiency and psychosocial distress secondary to dental features related to it. For all patients, regular dental check-ups and prevention of tooth decay in the form of oral hygiene instruction, dietary advice and appropriate use of fluoride is essential.

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