Dentinogenesis imperfecta associated with hearing Loss syndrome

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Abstract:

A four year old female patient presented to the department of Paediatric dentistry in Prince Zaid Bin Al - Hussein Hospital one of a military hospital in Jordan in June 2005 complaining of brownish opalescent discoloration of her the primary teeth.

This female child is found to have reddish brown to gray opalescent color enamel of primary teeth with breaks away enamel from the incisal edge of the anterior teeth and occlusal surface of posterior teeth with premature exfoliation of the primary teeth. The ENT findings are summarized by normal tympanic membrane on autoscopy and sensorineural hearing loss on pure tone eudiometry.

Keyword: Dentinogenesis imperfecta, discoloration, opalescent, hearing loss.

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Introduction:

Dentinogenesis imperfecta (DI) is a hereditary defect with autosomal recessive inheritance [1]. DI is the most common disorder affecting the structure of the dentin [2]. It is a genetic condition of mesodermal derivatives presenting with similar clinical and radiographic features in both the deciduous and permanent dentition. The estimated incidence is 1:8000 [2]. It has three types: type 1 DI has been defined as the dental defects associated with some forms of osteogenesis imperfecta (OI). Type II DI occurs as an isolated trait of dentinogenesis imperfecta [3]; type III, found only in a tri-racial inbred population in southern Maryland[4]. The disease consists of: delayed teeth eruption, mild mental retardation, short stature, sensorineural hearing loss and dysmorphic facies[2].

Histologically, the enamel is reported to be normal [5]; although hypo calcified defects may be present [3]. Enamel cracking appears within the enamel itself [6] or along the dentin-enamel junction[7], although the dentin-enamel junction appears qualitatively normal [6]. The mantle dentin has a normal tubular structure [6]. The cementum appears normal in structure and quantity [6].

Biochemically, DI-II may not be a collagen defect but a disorder of dentin mineralization [8, 9]. The mineral content is decreased, resulting from fewer crystals and increased water content [10].

A search of the literature came up with a few other DI-associated syndromes and there modes of inheritance. DI is the most common genetic disease of dentition. Actually it is the most common autosomal dominant disorder affecting humankind [11, 12]. Several syndromes are associated with dental findings which are clinically and
radio-graphically similar to those observed in DI-II. When conditions are found to be associated with DI, it is crucial to question if it is really DI or some phenocopy. Making the diagnosis of DI is also worth discussing dentin dysplasia type II looks similar or, sometimes, identical to DI in the primary dentition and may mislead some clinicians [13]. Discoloration of teeth appears to be the minimum criteria for clinicians in making diagnosis. Having denticles, pulp obliteration, and bulbous-shaped molars, but absence of tooth discoloration, does not fulfill the diagnostic criteria. It appears that the radiographic features are overruled by the clinical ones. Clinically normal teeth form patients with osteogenesis imperfecta have also been reported to have dentin aberrations [14]. The absence of discoloration or opalescence of teeth does not mean dentin is unaffected. From the molecular genetics and biochemical points of view, discoloration of the teeth should not be minimal criteria for making diagnosis of DI [15].

**Case report:**

A four year old female patient presented to the Paediatric dentistry clinic department in Price Zaid Ben Al-Hussien Hospital one of a military hospital in Jordan in June 2005 complaining of discoloration and wear of her teeth. Intra-oral examination revealed that reddish brown to gray opalescent color enamel of the primary teeth with breaks away enamel from the incisal edge of the anterior teeth and occlusal surface of the posterior teeth with premature exfoliation of the primary tooth (lower left primary central incisor) (Fig1). There was no soft tissue pathology with the exception of asymptomatic fistula located at the apical region of the upper right primary lateral incisor.
Panoramic radiograph shows the presence of all unerupted permanent teeth. The primary teeth had slender roots, bulbous crown, and pulp chamber entirely absent in primary teeth, pulp canals of the lower teeth are small and ribbon like (Fig 2). The tooth germ of the first permanent molars showed bulbous crowns with apparently normal enamel but constricted roots. The ENT findings are: normal tympanic membrane and high frequency sensorineural hearing loss associated with mild mental retardation and short stature.

The review of other systems is normal and the patient has a good general condition. The family history supported a provisional diagnosis of DI with a pedigree covering three generations. Consultation with Pediatricians revealed no evidence of ostogenesis imperfecta, such as a history of multiple fractures of her bones, laxity of the joints, or blue sclera.

**Management (of dental problems):**

Teeth affected by DI, especially in the primary dentition, require early treatment as this may intercept attrition and preserve function, aesthetics and normal growth [16]. Early treatment is necessary in dentinogenesis imperfecta as this can prevent pulpal pathosis. Two different modalities of treatment have been reported: complete restoration of the dentition with stainless steel crowns for the primary molars and composite crowns for the incisors [16],[17] or treatment with overlay dentures resting over the remnants of primary teeth [18],[19]. Difficulties may arise from decreased compliance with very young children making treatment under general anaesthesia usually necessary. Overdentures may cause hygiene problems provided by plaque retention gaps. Several methods of treating dentinogenesis imperfecta have reported successful, but long-term follow up is necessary to determine the ultimate
prognosis for each approach. Strict oral hygiene and adjustment of the restorations to changes in the stomatognathic system is the main points addressed during follow-up.

**Figure 1.** Generalized opalescent tooth discolorations, wear of the enamel were the clinical manifestation of dentinogenesis imperfecta.

**Figure 2.** Panoramic radiograph shows the wear of the deciduous teeth. Bulbous crowns, long and tapered roots, pulp obliteration of permanent dentition.
References
