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CASE REPORT

DENTINOGENESIS IMPERFECTA WITH OSTEOGENESIS IMPERFECTA: A CASE REPORT

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ABSTRACT: Dentinogegesis imperfecta (DI) represents a group of hereditary conditions that are characterized by abnormal dentin formation. These conditions are genetically and clinically heterogenous and can affect only the teeth or can be associated with the condition Osteogegesis imperfecta. The Osteogegesis imperfecta (OI) or the disease of fragile bones is a hereditary pathology affecting different tissues especially the bone. The teeth of DI cases wear more easily and excessively and also more susceptible to dental caries compared to normal teeth. Early prosthodontic rehabilitation can prevent or delay the wear as well as loss of teeth in DI. Herewith, we present case report of 10yr old boy with discolored, severely attrited permanent teeth with sinus openings. Along with systemic abnormalities like blue sclera, bow legs, protruded sternum. The case was diagnosed as Dentinogegesis imperfecta type I and discussed in this case report.

KEYWORDS: Opalescent dentin, Fragile bones, Collagen type I, Prosthodontic rehabilitation, Attrited.

INTRODUCTION: Dentinogegesis imperfecta (DI) or capdepont's teeth is a hereditary developmental disturbance of dentin.Dental changes seen in conjunction with Osteogegesis imperfecta, the systemic hereditary disorder of bone, is termed as Dentinogegesis imperfecta Type I.[1,2]

Osteogegesis imperfecta, type I-IV is an inherited disorder of the connective tissue formation. This has been associated with mutation of the COLI A1 and COLI A2 genes that encodes production of especially type I collagen, found in the bones, organ capsules, fascia, cornea, sclera, tendons, meninges, and dermis.In contrast DI is associated with mutation of the Dentin sialophosphoprotein gene (DSPP).Clinical features of Osteogegesis imperfecta are bone fragility, high frequency of fractures, bone deformity, joint hypermobility or hypomotility, subnormal or short stature, Dentinogegesis imperfecta (DI), bluish/greyish hue of sclera, hearing loss in adulthood, vascular, neurological and pulmonary complications.[1,2,3,4]

Because Osteogegesis imperfecta (OI) is a genetic generalized condition, it has no cure. Surgical correction of deformities, physiotherapy, use of orthotic support and devices to assist mobility (eg, wheelchairs) were the primary means of treatment. Bisphosphonates (eg, pamidronate) are synthetic analogues of pyrophosphate that inhibit osteoclast-mediated bone resorption on the endosteal surface of bone by binding to hydroxyapatite. As a result, unopposed osteoblastic new bone formation on the periosteal surface results in an increase in cortical thickness.[5]

DI is a localized mesodermal dysplasia affecting both the primary and permanent dentitions. The clinical features includes variation in color of the teeth from brown to blue, sometimes described as amber or gray, with an opalescent sheen. The enamel may show hypoplastic or hypocalcified defects in about one-third of the patients and tends to crack away from the defective dentin. The exposed dentin may undergo severe and rapid attrition.[6]Radiographically, the teeth have bulbous crowns with constricted short roots wide pulp chambers and resemble "shell teeth," but they will

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progressively obliterate. Histologically, the enamel, although normal in structure, tends to crack. The dentino-enamel junction is scalloped. The mantle dentin is normal, whereas the dentinal tubules of the circumferential dentin are coarse and branched and the total number of tubules is reduced. The presence of an atubular area in the dentin with reduced mineralization and a reduced number of odontoblasts are consistent findings. Pulpal inclusions and much interglobular dentin are also frequent. The biochemical characteristics of the dentin include a collagen defect and a primary defect in the calcifying matrix.[7,8]

Early diagnosis and treatment of both dentitions in DItype I is recommended, as it may prevent or intercept deterioration of the teeth, occlusion and improve esthetics. Further maintenance of deciduous teeth is therefore, particularly important to insure normal alignment of permanent teeth and to reduce the extensive orthodontic treatment.[9]

CASE REPORT: A 14-year-old male patient came to the Outpatient Department of Government Dental College and Hospital, RIMS, Kadapa, A.P. with a chief complaint of a pain in the lower front teeth for a week. The pain was severe, intermittent, and lasts for few minutes. It aggravates while chewing, on taking cold and hot food substances and relieves on medication. Family history revealed that he was third child of the parents married consanguineously. The other two children were found to be absolutely normal. The child was conscious and cooperative. His medical history revealed that he had a defective vision and hearing from birth. The Extra oral examination revealed decreased vertical height, facial symmetry with a concave profile and a class III skeletal pattern (Fig No. 1, 2). An intraoral examination revealed that the teeth present in the oral cavity were, 11, 12, 13, 14, 15, 16, 21, 22, 23, 24, 25, 26, 31, 32, 33, 34, 35, and 41, 42, 43, 44, 45. Teeth found to be missing were 17, 27, 36, 37, 46 and 47. 11, 15, 16, 21, 22, 24, 25, 31, 32, 33, 34, 41, 42, 43, and 45 were brownish in color. 12, 13, 14, 23, 35, and 44 showed opalescent translucent hue. Severe attrition seen in relation to 11, 12, 14, 15, 16, 21, 2, 23, 24, 25, 33, 36, 44, and 45. Dental caries in relation to 11, and 12.Sinus openings seen in relation to 31, 32, 33, 41, and 42. (Fig No. 5, 6, 7).

Panoramic radiograph showed that the few teeth had decreased thickness and a few with no enamel covering around the tooth crowns, large wide open pulp chambers with lake of pulp horns. Permanent teeth in relation to 17, 18, 27, 28, 36, 37, 46 and 47 were found absent. Developing teeth crowns 38, 48 seen. (Fig No. 8) A provisional diagnosis of Dentinogegesis imperfecta was made with the clinical and radiographic features.

The patient was sent to the pediatrician and on general physical examination found that he had limited mobility, bow legs, protuberance of sternum and defective hearing. No associated abnormalities of other systemic organs were found. The pediatrician was of the opinion that these features represent mild form of Osteogegesis imperfecta. So, the diagnosis of Dentinogegesis imperfecta Type I was confirmed after consulting the pediatrician.

The patient was advised to consult the Opthalmologist and ENT surgeon for treatment of visual and hearing defects.

As for as Dental findings, the management was planned as follows:

1. Antibiotics and anesthetics given to resolve infection and pain.
2. Extraction of 11,21 31,32, 41and 42.
3. Fixed partial dentures in relation to maxillary and mandibular teeth except mandibular molarseither ceramic or stainless steel if the teeth condition permits.

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4. Removable partial denture for missing mandibular posterior teeth.
5. Overdentures if the teeth cannot be fit for fixed prosthesis. Medication was given to the patient and asked him to come back after a week for further treatment. But, the patient never turned up.

DISCUSSION: The treatment in this case was planned assessing his oral condition that extraction of teeth with the sinus openings after controlling the infection and Prosthodontic rehabilitation of the remaining teeth and missing teeth. The aims of dental treatment for children with DI associated with OI are to ensure favorable conditions for eruption of the permanent teeth and normal growth of the facial bones and temporomandibular joints.10. The objectives of early treatment of DI in the primary dentition as follows.[9]

1. Maintain dental health and preserve vitality, form, and size of the dentition.
2. Provide the patient with an esthetic appearance at an early age, in order to prevent psychological problems.
3. Provide the patient with a functional dentition.
4. Prevent loss of vertical dimension, and maintain arch length.
5. Establish a rapport with the patient and the patient's family, yearly in the treatment.
6. Caries prevention, attrition observation, and monitoring of skeletal development.

Children diagnosed as having OI and also familial history of DI should be seen by a dentist as soon as possible after the eruption of the deciduous anterior teeth in order to determine whether there is DI involvement. The clinical severity of DI needs to be assessed when developing a treatment plan. When the disease is more severe, the biochemical properties of the enamel and dentin are compromised, and this can affect adhesion strength and can be treated only with complete dentures.

Shabtaisapir and Joseph saphir presents a case report where treatment done to a child diagnosed with DI at 20 months age for preventing the primary teeth from attrition and premature loss.[9] The sixmonth follow up examination revealed the restorations to be esthetically acceptable and functional. Thus, they recommend for severe cases of DI two treatment stages performed under general anesthesia. Stage 1 is early (around age 18-20 months) and is directed to cover the incisors with composite restorations and the first primary molars with preformed crowns. Stage 2 (around age 28-30 months) seeks to protect the second primary molars with preformed crowns and cover the canines with composite restorations.

Anil goud, Saredeshpande in a case report 'Prosthodontic rehabilitation of DI' suggests many treatment modalities like over dentures, stainless steel crowns, jacket crowns, pin-retained cast gold "thimbles" under acrylic resin crowns, stainless steel crowns with acrylic facing, and simple, removable appliances.[10]A combination of partial dentures and prosthetic crowns on the anterior teeth has also been described. Indirect resin crown (IRC) technique to restore the maxillary incisors has been used to treat a patient with severe enamel hypoplasia and attrition. This technique may offer esthetic, affordable, and long-lasting intermediate restoration. Orthodontic treatment has been successfully performed in patients with different degrees of DI. In less severe cases of DI, carbamide peroxide bleaching has been successfully used to treat discoloration.[10]

If the deciduous teeth begin to wear, placement of artificial crowns is recommended before excessive loss of tooth structure occurs.8In the restorative treatment of pediatric patients,

glassionomer with fluoride-releasing and chemically attaching materials are recommended for occlussally non-stressed areas. An acid etch technique followed by composite restoration is proposed as an alternative for restoration of the anterior teeth.

In the present case the patient reported for treatment of the toothache with gross destruction of permanent teeth. Medication was given for sibsiding the infection. Though we have explained him the prosthodontic treatment for functional and esthetic improvement after controlling infection and extraction of affected anterior teeth, the patient never turned up. Thus this case stresses the importance to bring the awareness in the public, the temporary and permanent treatment available in these types of genetically altered teeth.

CONCLUSION: Early diagnosis and treatment of DI with OI is recommended, as it may prevent or intercept deterioration of the teeth, occlusion and improve esthetics. Minimum treatment modalities are present for Osteogegesis imperfecta as discussed above because it is a generalized disease. But Dentinogegesis imperfecta can be repairable to improve the functional and esthetics' of dentition with the present available dental treatments. The present case necessasites us to bring awareness about these types of diseases in the public through various media to educate and motivate the patients for consulting the dentist as soon as the deciduous and permanent teeth erupt, so that an attempt can be made to prevent loss of tooth structure.

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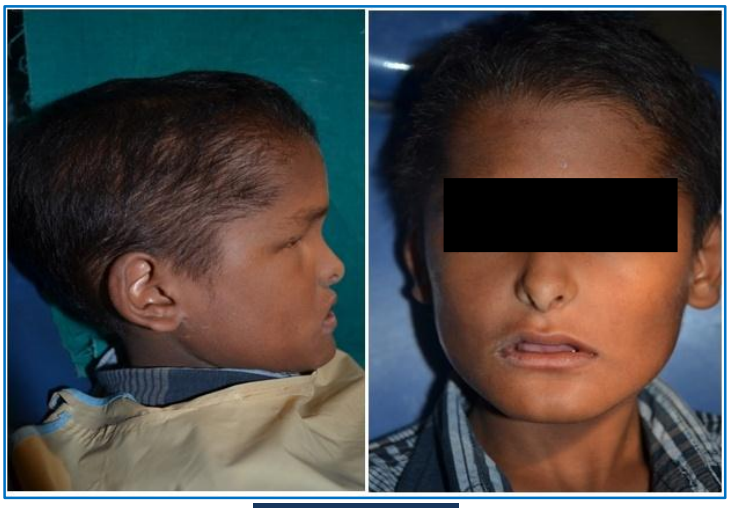

Fig. 1 & 2

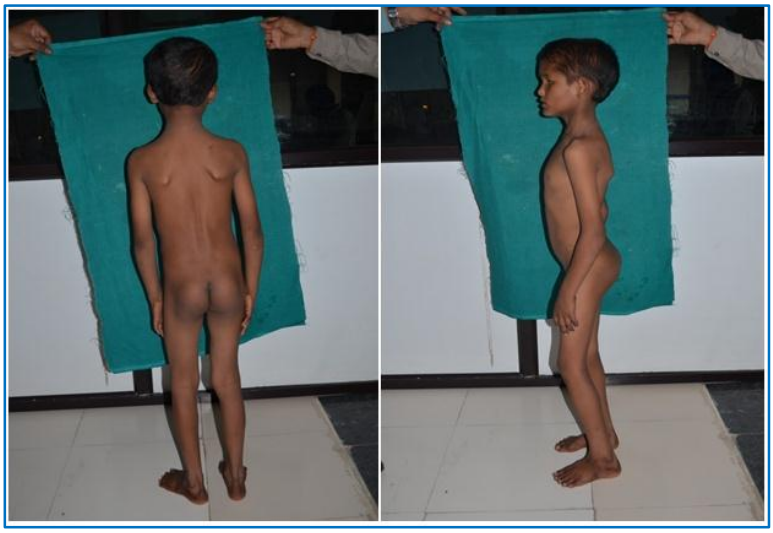

Fig. 3 & 4

Fig. 5 & 6

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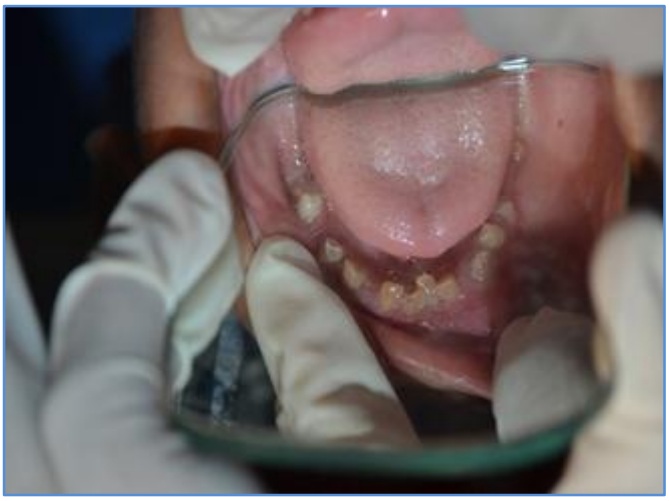

Fig. 7

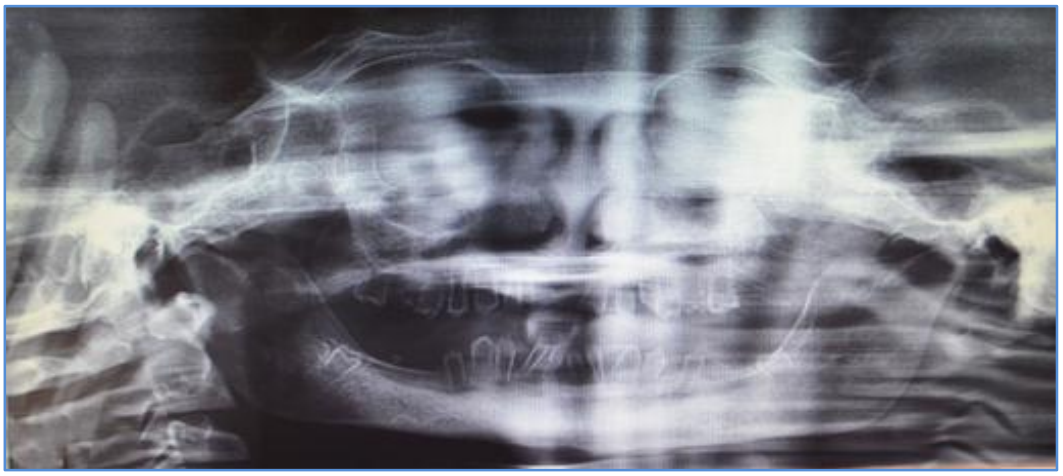

Fig. 8

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