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Dentinogenesis Imperfecta Type I - A Report of Two Cases and A Short Review of Literature

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Case Report

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Abstract: Dentinogenesis Imperfecta (DI) is a hereditary disorder affecting both the deciduous and permanent dentition. This disorder classically affects the dentin and it is characterized by the presence of opalescent dentin caused due to autosomal dominant pattern resulting in a dusky blue to brownish discoloration of the teeth. It is classified as DI-I and DI-II. This present article discusses two case reports of Dentinogenesis Imperfecta. First is a case of 29-year-old female patient who reported to the dental O.P for an aesthetic correction. Second is a case of 19- year old female patient who reported to the dental O.P for an orthodontic correction.
Keywords: Dentinogenesis imperfecta, Autosomal dominant, Hereditary, Capdepont, Opalescent dentin, Pulpal obliteration.

INTRODUCTION

Dentinogenesis Imperfecta is commonly known as hereditary opalescent dentin or Capdepont dysplasia [1]. It is a localized mesodermal dysplasia affecting both primary and permanent dentition and is a genetic disorder of tooth development inherited in an autosomal dominant pattern caused by mutations in the Dentin Sialo Phosphoprotein (DSPP) gene (cytogenetic location of 4q21.3). The term 'dentinogenesis imperfecta' was coined by Robert and Schour in 1939 and probably first recognized by Barret in1882 [2]. The frequency of occurrence of this genetic dental disorder is 1 in 6000-8000 births [1].

CASE REPORT 1

A 29 year old female patient reported to the department of Oral Medicine and Radiology with the chief complaint of Bluish grey stains on all the surfaces of her teeth since childhood.

On obtaining a detailed family history it was found that she was the first child among the family of three siblings born to a non consanguineous parents, patient gave a history that her father, younger brother and sister had similar shade of teeth as that of her's ,whereas she claimed that her mother had a normal shade of teeth. Patient was married and has a six year old son who also has similar blue shade of teeth. She reported no history of bone fragility or bone fractures in her family.

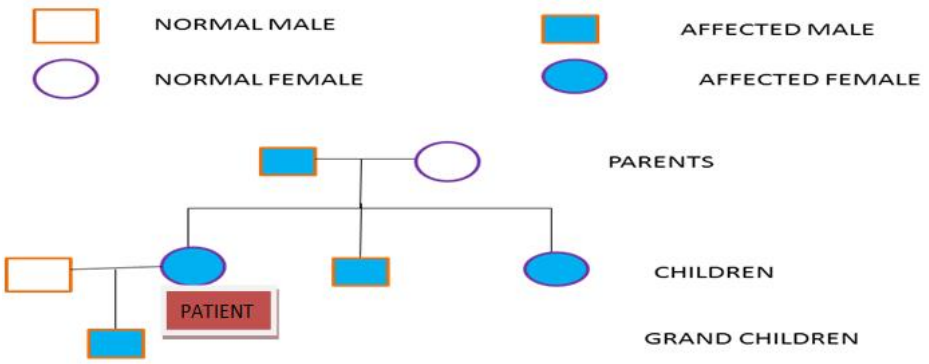

Fig-1: Pedigree diagram showing the mode of inheritance of case 1

Our patient was depressed as she was unable to speak, smile or socialize with others because of the abnormal shade of her teeth (Figure-2). Patient had no contributory past medical / surgical history. Patient gave a history of extraction of her decayed left and right lower molar teeth two years back.

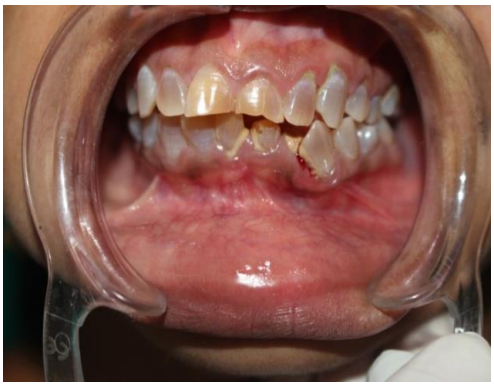

Fig-2: Generalized yellowish brown and translucent teeth

On Clinical examination there was presence of generalized bluish grey opalescent hue on labial and buccal surfaces of anterior and posterior teeth in relation to the maxillary and mandibular arch. Yellowish discoloration with loss of the enamel at the incisal third of 11 and 21(Figure-3) (c) was evident and significant attrition was observed in the mandibular anteriors .Patient gave a positive history of enamel getting chipped occasionally. Patient had dental caries in relation to left lower first molar and right lower third molar , root stump was evident in right upper third molar. The colour, size, texture, and contour of the maxillary and mandibular gingiva were within normal limits. Mild generalized plaque and calculus deposits were found mostly on the mandibular anterior and maxillary posterior teeth .There were no skeletal or scleral abnormalities.

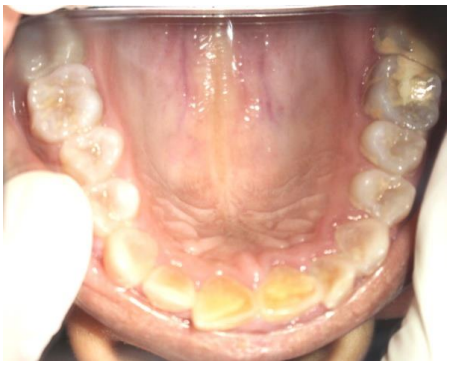

Figure-3 (a)

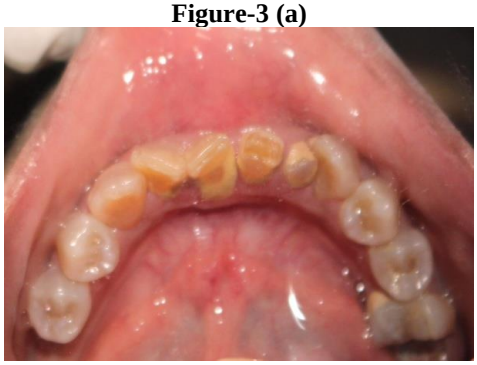

Figure-3 (b)

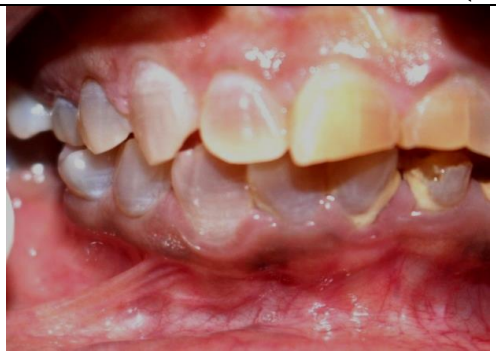

Figure-3 (c)

Fig-3 (a): Maxillary arch. (b) Mandibular arch (c) Right occlusion in maximal intercuspal position

Patient was subjected to intra oral periapical radiographs in relation to, maxillary anterior and mandibular anterior region. The IOPA in relation to the maxillary anterior region (Figure-4) revealed significantly shorter size of the teeth with loss of enamel structure at the incisal third, there was no significant difference in the radiodensities of enamel and dentin and there was a faint line of root canal at the level of apical third of the roots in relation to 11,12,13 and 21 .

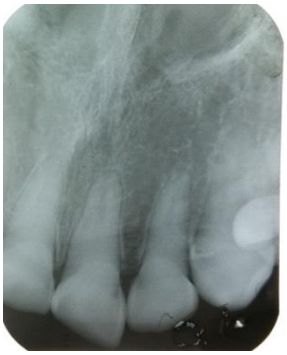

Fig-4: Iopa of maxillary anteriors

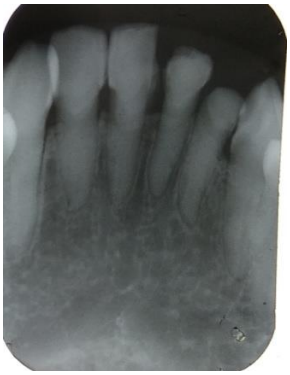

Fig-5: Iopa of mandibular anteriors

The IOPA in relation to the mandibular anterior region (Figure-5) revealed significantly short size of the teeth with loss of enamel structure at the incisal third in 31, incisal and middle third of the crown in relation to 32. In relation to 41, 42, 31 and 32 there was proximal loss of enamel structure leaving behind the open contact in between the teeth were evident. There was no significant difference in the radio densities of enamel and dentin and there was complete obliteration of pulp chamber in relation to all teeth.

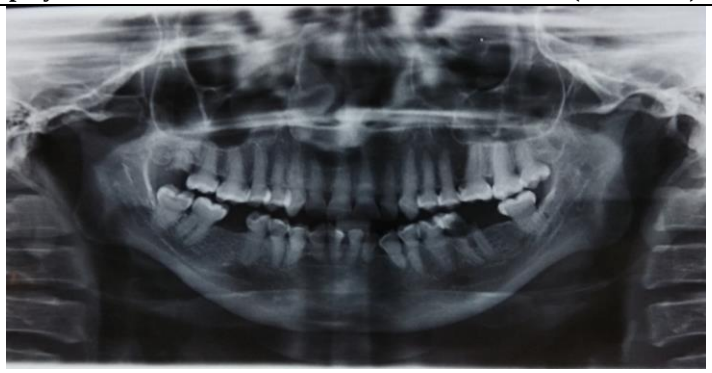

Fig-6: orthopantamogram

Further patient was subjected Orthopantamogram (Figure-6) which revealed that the trabecular bone pattern was normal. However, the bone appeared to be denser than normal. Note that bone in DI commonly looks denser because the observer's eye is used to contrast it with normal dentin. The crown to root ratio ranged from 1:2 to 1:3. All the teeth appeared to have bulbous crowns with a constricted cervical area. The remaining enamel appeared to have normal radiographic density with varying degrees of pulpal obliteration because of dentin hypertrophy. Bilaterally altered morphology in the head of the condyles were evident suggestive of early erosion /osteoarthritic changes of TMJ but the patient never exhibited any symptoms when TMJ examination was done. Absence of 37, 46 and root stumps in relation to 18 was present .The clinical and radiographic findings supported a diagnosis of Dentinogenesis Imperfecta type I (Revised classification) which coincides with the Dentinogenesis Imperfecta type II of Sheild's Classification.

TREATMENT PLAN

The following treatment plan was suggested in detail before the commencement of the treatment,. Risks, alternatives, and benefits of treatment were explained to the patient. Oral prophylaxis was done following which electric pulp testing was done in all the incisors and premolars. The anterior teeth and premolars were planned to be restored with bonded lithium disilicated glass-ceramic pressed veneers and crowns, root canal treatment was for 32 and extraction of grossly decayed 36, root stumps in 18, and 28 was advised. This case demonstrates that restoring functional occlusion and aesthetics for a patient with DI can be completed successfully using contemporary implant therapy and adhesive dentistry.

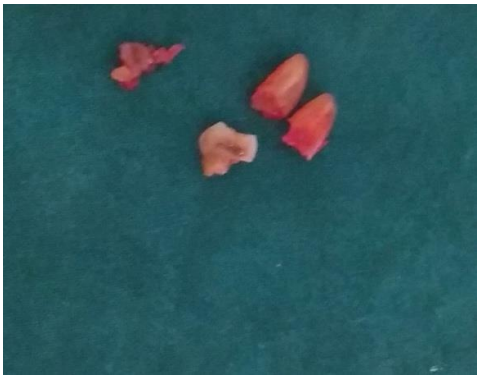

Fig-7: Extracted tooth fragments of 36 and 18

Fig-8: Extracted tooth – 28

Histopathological examination

The extracted root stumps 18 and grossly decayed 36 was submitted for histopathological examination and ground section (Figure-9) exhibited haphazardly arranged dentinal tubules, atubular dentin , large amount of cellular cementum with lack of dentino enamel junction in many areas suggestive of Dentinogenesis imperfecta (Figure-10).

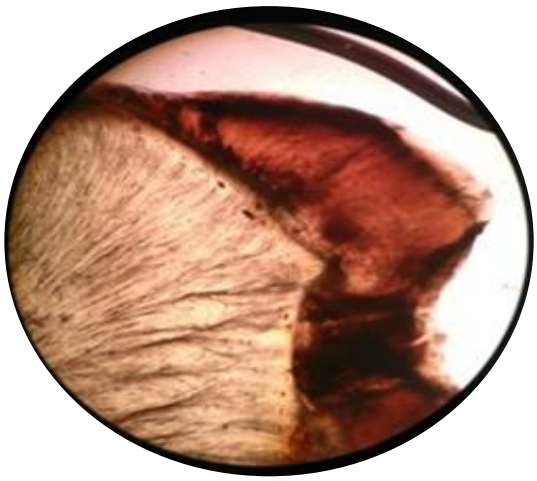

Fig-9: Ground section

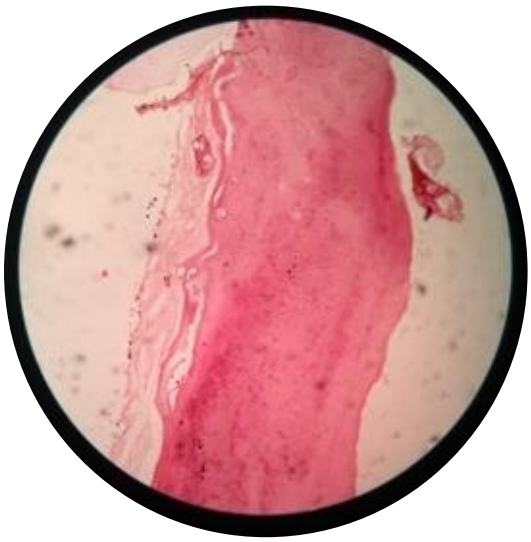

Fig-10: Histopathological section

CASE REPORT 2

A 19 year old female patient (Figure-11) reported to the department of Oral Medicine and Radiology with the chief complaint of irregularly placed upper and lower front teeth (Figure-12)for the past 8 years and wanted to get her teeth aligned. Patients past surgical history, she revealed that she underwent a cardiac surgery for the closure of patent ductus arteriosus, three years ago. Her past dental history revealed that she underwent a oral prophylaxis and extraction of her decayed teeth two years ago. She gave a history that some of her teeth had a peculiar yellowish brown shade, when questioned about her family history she revealed that she was second child among four siblings born to non consanguineous parents. She revealed that her father and her younger sister had similar shade of the teeth. There were no skeletal or scleral abnormalities and no familial history of bone fractures.

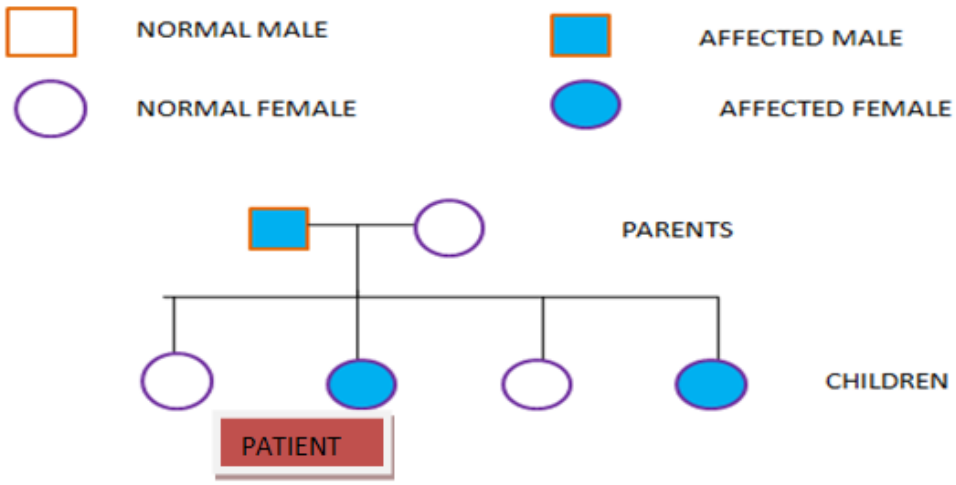

Fig-11: Pedigree diagram showing the mode of inheritance of case 2

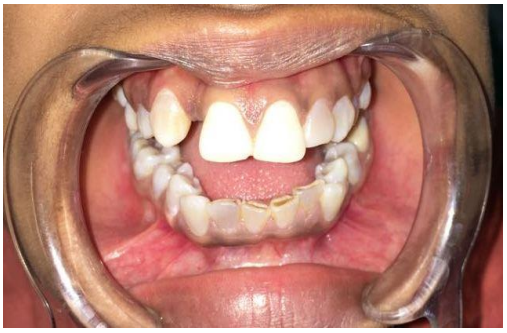

Fig-12: Intra oral photograph

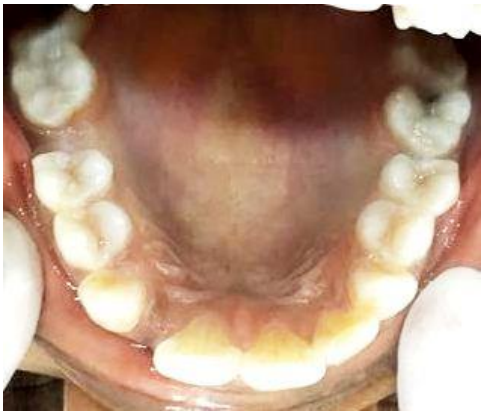

Fig-13: Maxillary arch

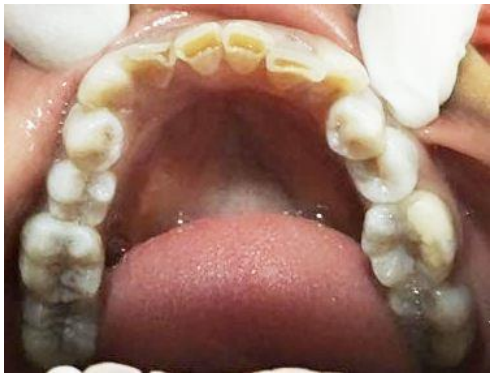

Fig-14: Mandibular arch

On Clinical examination patient had edge to edge bite with class I malocclusion. There was presence of generalized bluish opalescent hue on labial, buccal surfaces of anterior and posterior teeth in relation to the maxillary and mandibular arch. (Figure-13 &14) Yellowish discoloration with loss of the enamel at the incisal third of 31, 32, 41 and 42 was evident and patient gave a positive history of enamel getting chipped occasionally, had missing teeth in relation to 12, 16 and 26 was evident dental caries with chronic pulpitis were evident in 27, restored tooth 36. Intra oral periapical radiographs were taken in relation to mandibular anteriors (Figure-15) and left maxillary posteriors (Figure-16).

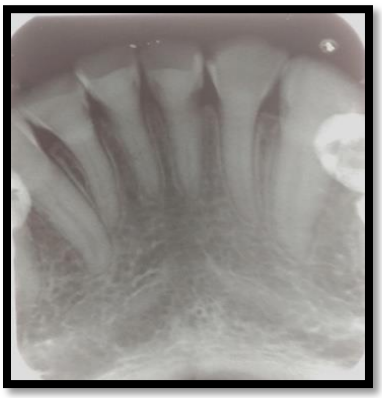

Fig-15: Iopa of mandibular anteriors

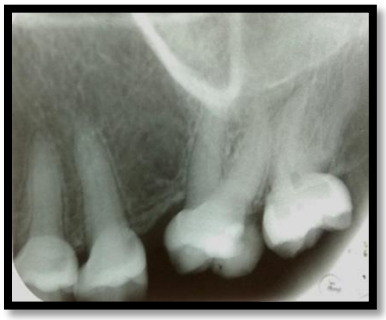

Fig-16: Iopa of left maxillary posteriors

The IOPA in relation to 24,25,26,27 and 28 region reveals all the teeth had a cervical constriction with bulbous crown; there was no marked difference in the radio density of dentin and complete obliteration of the pulp chamber. There was missing 26, in relation to 27 there was mesial migration of 27 towards the edentulous space and dorsoproximal radiolucency evident in the crown involving enamel, dentin suggestive of dental caries in mesial migrated 27.

The IOPA in relation to the mandibular anterior region revealed significantly shorter size of the teeth with loss of enamel structure at the incisal third in 31,32,41 and 42 There was no significant difference in the radio densities of enamel and dentin and there was faint radiolucency of root canals of 31,32,33 and 41,42 43.

OPG (Figure-17) revealed presence 29 teeth totally. In relation to 12,16,26 there was complete loss of tooth structure suggestive of missing teeth, The crowns of all the premolars and molars had a characteristic peculiar shape resembling that of a "Tulip" flower. There was a marked constriction of the crowns at cervical area. The maxillary and mandibular posterior teeth had complete obliteration of the pulp canals. In relation to 38 and 48 there was ¾ th of the root completion evident. The clinical and radiographic findings supported a diagnosis of Dentinogenesis Imperfecta typeI (Revised classification) which coincides with the Dentinogenesis Imperfecta type II of Sheild's Classification
(Figure-17)

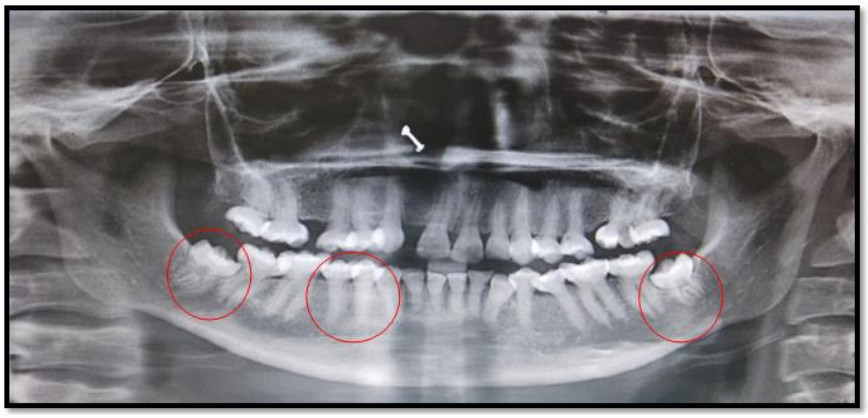

Fig-17: Orthopantamogram

TREATMENT PLAN

Patient is under Orthodontic treatment.

DISCUSSION

Dentinogenesis imperfecta (DI) is one of the most common genetic disorders affecting the structure of dentin[1,3]. The classification system for DI was proposed by Shield as - type 1, type 2 and type3. However, shield's classification had certain drawbacks, so it was revised into DI-I and DI-II. Some researchers believe that DI types II and III, and dentin dysplasia type II (which is also a hereditary dentin defect) are actually variant forms of the same disorder[3].

Various classifications proposed by different authors

Shields	Witkop	REvised	Clinical presentation
DI-I	DI	No substitute	Osteogenesis imperfecta with opalescent teeth
DI-II	Hereditary Opalescent Teeth	DI-I	Isolated opalescent teeth
DI-III	Brandywine isolate-found only in a population of southern Maryland (USA)	DI-II	Isolated opalescent teeth

DI: Dentinogenesis imperfecta

Formation of defective dentin in the structure and composition are the major anomalies of DI [8-12]. The enamel is usually reported to be normal, but is easy to chip or shear away from the dentin due to a lack of support from a normal scalloped dentin-enamel junction [8,11,12]. The early loss of enamel and poorly calcified dentin make these teeth prone to rapid wear. Teeth that undergo severe attrition may lead to pulp exposure and require endodontic treatment. However narrowing of the root canal and the defective dentin structure present a challenge for root canal treatment. In DI-affected teeth, the micro hardness of the dentin closely approximates that of cementum [9]. However DI-affected dentin has irregular tubules, often with larger areas ofuncalcified matrix, and contains up to 60% more water than do tubules in normal dentin [10-12].

Histologically the enamel, although normal in structure, tends to crack. The dentin-enamel junction is not scalloped. In most cases the structure of the mantle dentin is normal [6,7] whereas the dentinal tubules of the circumferential dentin are coarse and branched and the total number of tubules is reduced[6,7]. All of which is present in the first case. The presence of and a tubular area in the dentin with reduced mineralization and a reduced number of odontoblasts are consistent findings [6, 7]. Pulpal inclusions and much inter globular dentin are also frequent.

There are no guidelines on restorative treatment in teeth affected by DI. The treatment should begin as early as possible, considering the degree of tooth destruction and patient cooperation [15,17]. Treatment involves removal of sources of infection or pain if present, improvement of aesthetics and protection of the posterior teeth from wear. Beginning in infancy, treatment usually continues into adulthood with a number of options including the use of crowns, overdentures and dental implants depending on the age of the patient and the condition of the dentition. If the diagnosis occurs early in life, treatment options are

ample with good prognosis[15,16]. The objectives of early treatment of DI in the primary

Dentition is as follows:

- Maintain dental health and preserve vitality, form, and size of the dentition.
- Provide the patient with an esthetic appearance at an early age, in order to prevent psychological problems.
- Provide the patient with a functional dentition.
- Prevent loss of vertical dimension.
- Maintain arch length.
- Avoid interfering with the eruption of the remaining permanent teeth.
- Allow normal growth of the facial bones and temporomandibular joint.
- Establish a rapport with the patient and the patient's family early in the treatment [19].

Treatment demands a multidisciplinary approach. Collaboration of the pediatric dentist with a prosthodontist and an orthodontist is often imperative. Although caries is not a major concern in most cases, strict oral hygiene instructions and preventive treatment is important in order to prevent caries from adding to existing problems. Frequently there is a need to re-establish the vertical dimension of occlusion in order to restore the occlusion in the mixed and permanent dentition. Prosthetic restoration combined with orthodontic treatment may be advantageous, and evaluation of the occlusion prior to initiation of treatment is advised [15,18].

CONCLUSION

Genetically inherited disorders impact not only the physical condition, but also the psychological and social health of patients and their families. Patients can be unaware about the hereditary nature of this condition but it is the sole duty and responsibility of the dentist to make them understand the unique aspects of disease and the available modalities of treatment.

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