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"PERFECTING THE IMPERFECTA" CASE REPORT ON AMELOGENESIS IMPERFECTA

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ABSTRACT
Amelogenesis Imperfecta delineates a rare group of developmental conditions, which are genomic in origin, affecting the clinical advent of the enamel and structure of all or nearly all the teeth in a more or less equal fashion, often in conjunction with morphologic or biochemical changes elsewhere in the body. The present article proffer insight into how to diagnose Amelogenesis imperfecta based on clinical and radiologic features and outline necessary treatment solutions for the management of the condition. Successful management led to improvement of function and aesthetics as well as quality of life.

Keywords: Amelogenesis, rehabilitation, hypomaturation, developmental, hypocalcified, occlusion.

INTRODUCTION

Amelogenesis imperfecta delineates a rare group of developmental conditions, which are genomic in origin, affecting the clinical advent of the enamel and structure of all or nearly all the teeth in a more or less equal fashion, often in conjunction with morphologic or biochemical changes elsewhere in the body.[1] It is due to the malfunction of the proteins in the enamel (ameloblastin, enamelin, tuftelin and amelogenin) caused by mutations or altered expression in five genes namely AMEL(amelogenin), ENAM (enamelin), MMP20 (matrix metalloproteinaise-20), KLK4 (kallikrein-4) and FAM83 resulting in abnormal enamel formation via amelogenesis.[2]

Enamel is an epithelial-derived tissue comprised of highly organised hydroxyapatite crystals that form in a defined extracellular space. It is acellular and has no intrinsic repair potential. Enamel formation is a highly orchestrated process initiated by ameloblasts during tooth developmental stages after dentinogenesis. The entire process is under genetic control. Diverse phenotypes showing a wide spectrum of characteristics result from genetic mutations.[3]

The reported prevalence of AI varies widely in the literature (1:700 to 1:14,000), according to the populations studied.[4] Literature offers a plethora of classification systems but the classification systems are based exclusively on phenotype and are perplexing. It is not always possible to know which classification system might have been applied to a particular case.

To clean the air, Aldred et al. discussed previous systems used and proposed a new model for

classifying AI based on four broad areas. These are described below:[5]

- Mode of inheritance: autosomal dominant, autosomal recessive, X-linked, isolated case.
- Molecular basis: chromosomal localisation/locus/mutation.
- Biochemical outcome: putative result of mutation when known.
- Phenotype: hypoplastic, hypocalcified, hypomaturation, hypomaturation-hypoplastic with taurodontism.

Clinical characteristics of **Hypoplastic AI** range from reduced enamel thickness, to pitting and hard or translucent surface. Radiographically, the enamel contrasts normally from dentine.

Whereas in **Hypocalcified AI,** there is defect in enamel calcification. Although weak in structure, thickness is normal. It appears opaque or chalky, stained and wears down rapidly. Radiographically, enamel is less radio-opaque than dentine.

In **Hypomaturation AI**, thickness of enamel is normal but it is mottled in appearance, vulnerable to tooth wear (but not as severe as the hypocalcified type). Radiographically, radiodensity is same as dentine.

Characteristics of hypomaturation-hypoplasia

- Mixed hypomaturation and hypoplasia appearance[6]

Other clinical features evident may be delay in dental eruption, microdontia, deviant crown and morphology, root resorption, short roots, enlarged pulp chamber, pulp stones, dens in dente, tooth agenesis, crowding of teeth, gingival enlargement, gingivitis and periodontitis. Additionally there are other reported skeletal abnormalities such as overbite, overjet and cross bites. [7]

The treatment planning for patients with amelogenesis imperfecta is related to many factors: the age, socioeconomic status of the patient, the type and severity of the disorder, and the intraoral condition.[8]

CASE REPORT

This clinical report describes the complete rehabilitation of Amelogenesis imperfecta in a young adult. An 18 year old female who is a native of Himachal Pradesh was referred for specialist dental care to the department of Conservative dentistry, Swami Devi Dyal Hospital and Dental College. The patient was aware and very self-conscious of her not so good looking teeth. From the family medical history it appeared that her grandfather was affected by a hereditary enamel defect. Medical history was unremarkable. This girl's parents did not seek any treatment previously, thinking that the condition was not resulting in any other systemic manifestations. Also, because of the financial constraints there was little they could really do about it and accepted it as part of her appearance. This case report describes the successful management of an unaesthetic smile due to Amelogenesis imperfecta.

EXAMINATION

A physical intraoral examination revealed that the teeth were found to possess very little enamel; crowns were paper thin and short. Yellowish discoloration of entire dentition was evident because of very thin or almost no enamel. The crown morphology of teeth was erratic and occlusal surfaces of anterior and exterior sections were rough and irregular. There was dental calculus at few proximal surfaces. There were defects of the crowns of the incisors involving the incisal and median thirds and an anterior open bite was present. She did not exhibit any periodontal problems. Attrition of the molars had resulted in a decrease of the vertical dimension of occlusion. The teeth exhibited microdontia with spacing between them. One of the central incisors was severely discolored. Her past medical history was noncontributory. The deciduous teeth had also been affected but only to a minor degree.

Volume 2, Issue 2; March-April 2019; Page No. 85-92

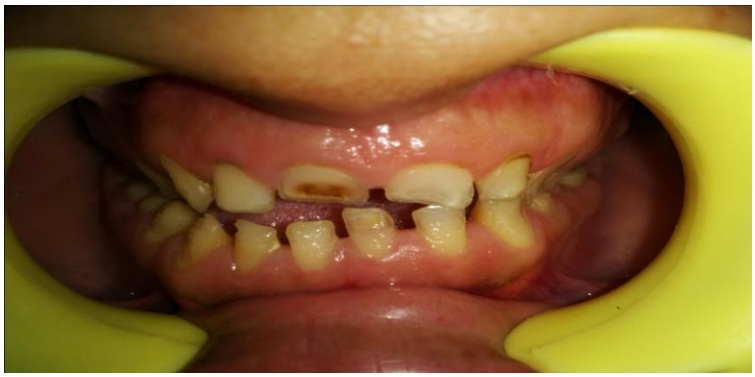

Figure 1 View of dentition before treatment

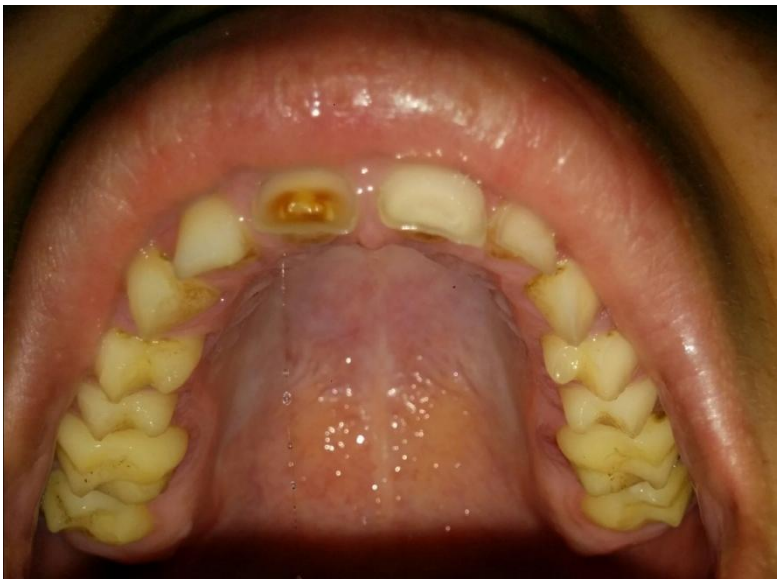

Figure 2 Occlusal view showing erratic occlusal surfaces (rough and irregular) and paper thin incisal edges

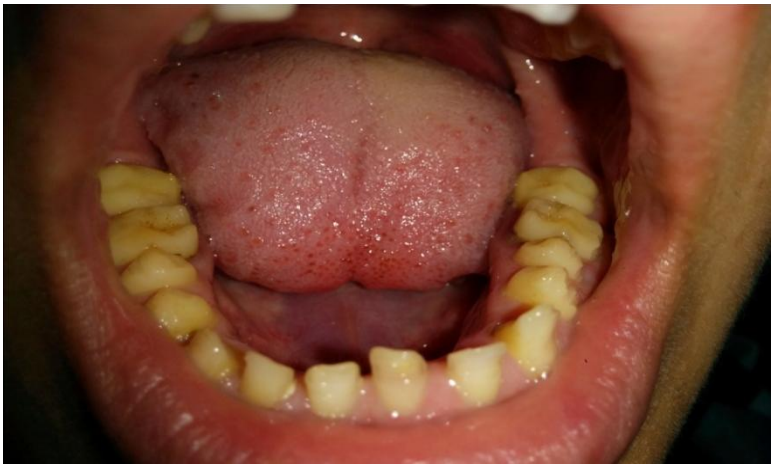

Figure 3 Occlusal view of the lower arch

Volume 2, Issue 2; March-April 2019; Page No. 85-92

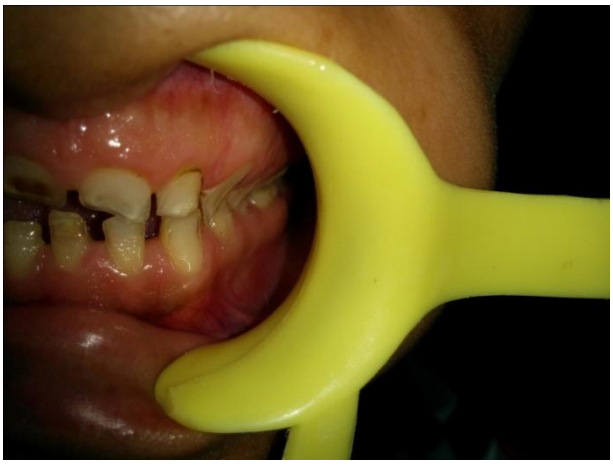

Figure 4 Left lateral view in MIP

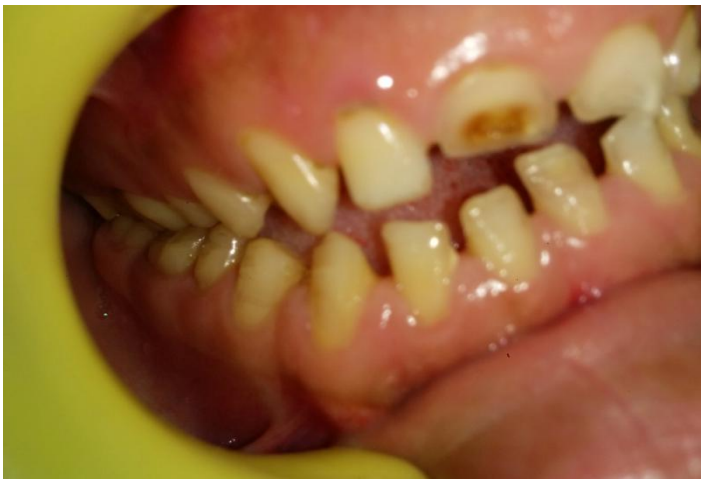

Figure 5 Right lateral views in maximum intercuspation

INVESTIGATIONS:

- Panoramic radiograph was advised. The patient's initial orthopantomograph showed presence of a thin layer of enamel with radiodensity of enamel more than dentin.

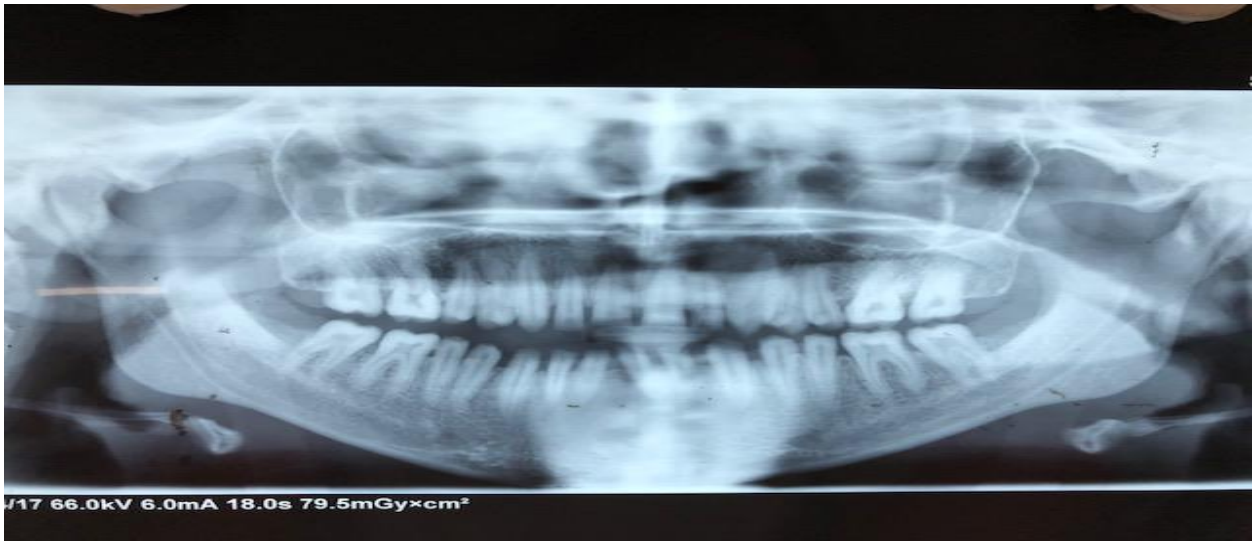

Figure 6 Panorex before treatment

Volume 2, Issue 2; March-April 2019; Page No. 85-92

- Vitality tests revealed that the teeth 11,12,13,21,22,23, 24, 31,41 were non vital as compared to adjacent/ contralateral teeth.
- A consultation with nephrologist was suggested as an association between AI and nephrocalcinosis is usually reported in literature. Hence, the report was negative for renal pathology but the patient was advised for regular supervision to detect any onset of the disease at a later stage of life.

DIAGNOSIS

Based on the clinical and radiographic examination final diagnosis of hereditary hypoplastic-hypomaturation-AI was made.

1) Generalized enamel hypoplasia of permanent dentition; (evident clinically and on radiograph)

2) Family history of the condition, although in the recessive forms.

3) Absence of systemic diseases that may cause generalized enamel hypoplasia resembling Al (e.g. systemic disorders involving calcium metabolism such as renal and liver disorders).

In addition, conditions which show hypocalcification enamel defects such as the trichodentoosseous (TOO) syndrome (kinky hair, dysplastic nails, sclerotic bones, enamel hypoplasia, severe taurodontism) were excluded. Variants of ectodermal dysplasia, which may also show generalized enamel hypoplasia, as well as fluorosis also were excluded.[9]

TREATMENT PLANNING

The supportive clinical care needed by these individuals is worthwhile both in terms of clinical and emotional demands.[10] Intervention will likely be earlier and more comprehensive than for others. Various treatment approaches have been described for rehabilitation of amelogenesis imperfecta in adults and children.[11] The use of composite resins in aesthetic restoration of permanent anterior and posterior teeth as affected by hypoplastic AI type was impossible for this case as patient suffered from more attrition as in hypomineralized varieties. No attempt was made to increase the vertical dimension of occlusion. The longer-term care still revolves around either crowns or, more frequently these days, adhesive, plastic restorations. Many dentists endeavor appositely to delay the first "tooth-cutting" restoration as conversations with a substantial number of people with AI suggest that this professional moderation may be unwelcome. Some of these same adults will recount that, if they had realized that restored teeth must eventually fail, they would have chosen tooth-tissue destructive, but aesthetically more appealing restorations earlier in their adolescence, in order to appear most "ordinary" to their peers at an important time in social development.[1]

Pulp vitality was maintained for molars. In the first phase of treatment endodontic therapy was carried out for teeth 15,14,13,12,11,21,22,23,24,25,31,32,33,34,35,41,42, 43,44,45. After access cavity preparation and working length determination, canals were prepared using NeoEndo rotary files (Orikam). Irrigation was done alternatively using Sodium Hypochlorite 1.3%, Saline, Chlorhexidine 2% and EDTA. Thereafter canals were dried with sterile paper points and were obturated with gutta percha and AH Plus sealer. Finally access cavity was filled.

Volume 2, Issue 2; March-April 2019; Page No. 85-92

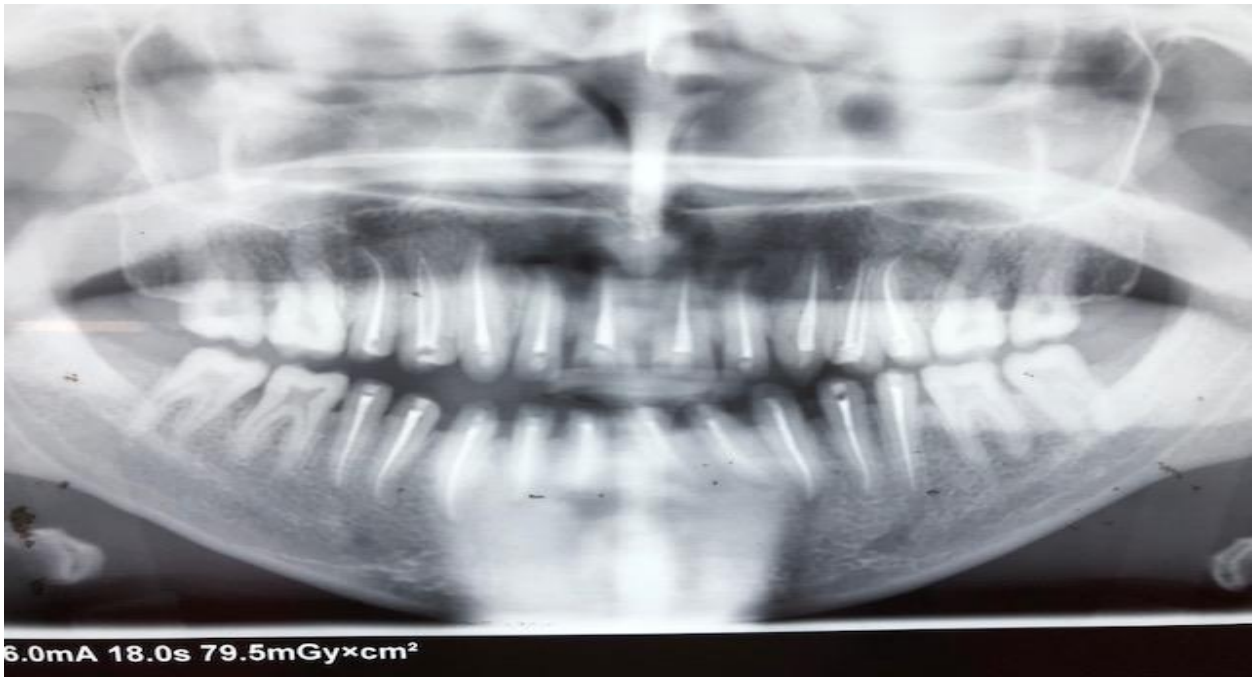

Figure 7 Panorex after endodontic treatment

Second phase consisted of tooth preparation for the same teeth to receive all ceramics crowns. The appropriate shade was then selected using the VITA shade guide (Vita Zahnfabrik, Badsackingen, Germany) prior to preparation. Gingival retraction was done for prepared teeth. Impressions were made with double mix, single impression technique using additional silicone (Dentsply Sirona, Aquasil) with light body (Aquasil Ultra LV) in a custom tray. Patient was guided into centric relation and bite registration was done using PVS bite registration paste (Jetbite, Coltene). The prepared teeth were restored with provisional crowns fabricated with 3M ESPE Protemp ™ 4, using putty indices of the diagnostic wax-up. Temporary crowns were cemented using temporary cement (3M ESPE, Rely X™ Temp NE). Face-bow transfer (Hanau spring-bow) was done to mount maxillary cast on a semi-adjustable articulator with the mandibular cast with the help of the bite registration record. All-ceramic crowns (IPS Emax), were fabricated and trial was done in the patients mouth. The marginal fit and esthetic appearance of the crowns and veneers were verified. Etching was done using Ultradent Porcelain Etch and Silane). In the final steps, the crowns were cemented using resin cement (Rely X Ultimate Clicker™, 3M ESPE). At the end of the procedure, we were able to achieve both functional and esthetic satisfaction of the patient. After a follow up of six months, the patient did not present any signs and symptoms of abnormal function and was satisfied with both function and esthetics.

Volume 2, Issue 2; March-April 2019; Page No. 85-92

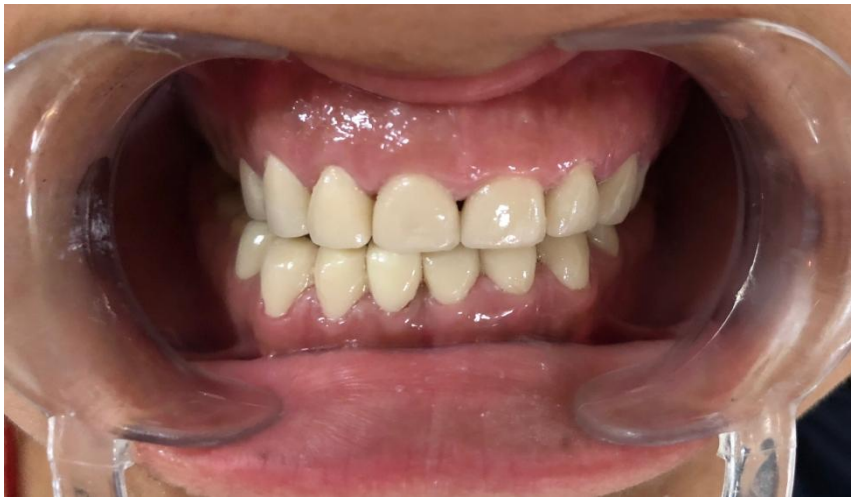

Figure 8 View of prostheses after fitting.

DIFFERENTIAL DIAGNOSIS

- Extrinsic disorders of tooth formation
- Chronological disorders of tooth formation
- Localised disorders of tooth formation should be considered in the differential diagnosis.
- The commonest differential diagnosis is dental fluorosis.
- Chronological enamel hypoplasia – can arise from one of many causes during the time of tooth formation as in cases of gastrointestinal disorders, such as coeliac disease, anti-leukaemic therapy.
- Molar-Incisor Hypomineralisation (MIH)[12]

Consideration should be given to

a) does anyone else in the family have anything like this;

b) are all of the teeth affected in a similar manner;

c) is there a chronological distribution to the appearance seen;

d) is there anything in the past medical history which might have caused sufficient metabolic disturbance to affect enamel formation – and form the foundation for differential diagnosis.

DISCUSSION

The appearance of teeth and problems associated with the enamel (such as sensitivity, staining and roughness) can be of major psychological and functional concern to the patient. The aim of any treatment plan is early diagnosis, prevention, stabilisation, restoration of any defects and regular maintenance.[13] The rehabilitation of amelogenesis imperfecta in an adult must take into account occlusion, periodontal health and esthetics, vitality status of teeth, appearance of teeth.[14] The use of composite resins in aesthetic restoration of permanent anterior and posterior teeth was impossible for this case as patient suffered from more attrition as in hypomineralized variant of AI. Patient was encouraged to opt for phased treatment including prosthodontic/ orthodontic treatment to raise the vertical dimension/ face length, periodontal procedures for crown lengthening and gum line correction and finally prosthodontic rehabilitation. But because of the financial concerns and personal choice, she unequivocally pitched forthwith final stage of treatment that is prosthodontic rehabilitation using all ceramic crowns. She was very well informed that the long survival of rehabilitation revolves around maintainence of oral hygiene.[15]

CONCLUSION:

This clinical report recounts the utilization of indirect all ceramic restorations to restore the masticatory function, improve the esthetics with careful consideration of patient expectations and taking into consideration financial constraints. In conclusion, patients affected by Amelogenesis imperfecta require prosthetic rehabilitation besides management of psychological condition on account of social stigma.

REFERENCES:

Volume 2, Issue 2; March-April 2019; Page No. 85-92

1. Crawford PJM, Aldred M, Zupan AB. Amelogenesis imperfect. Orphanet J Rare Dis. 2007; 17(2):1-23.
2. Gadhia K, McDonald S, Arkutu N, Malik K. Amelogenesis imperfecta: an introduction. BDJ. 2012;12 (2), 377–379.
3. Lacruz RS, Habelitz S, Wright JT, Paine ML. Dental Enamel Formation and Implications for Oral Health and Disease. Physiol Rev. 2017; 97(3): 939–993.
4. Santos, Maria Cristina Leme Godoy dos, & Line, Sergio Roberto Peres. The genetics of amelogenesis imperfecta: a review of the literature. J Appl Oral Sci. 2005;13(3), 212-217.
5. Aldred MJ, Crawford PJ. Amelogenesis imperfecta—towards a new classification. Oral Dis. 1995;1(1):2-5.
6. Seow WK. Clinical diagnosis and management srategies of amelogenesis imperfecta variants. Pediatric Dentistry.1993;15(6).
7. Poulsen S, Gjørup H, Haubek D, Haukali G, Hintze H, Løvschall H & Errboe M. Amelogenesis imperfecta – a systematic literature review of associated dental and oro-facial abnormalities and their impact on patients. Acta Odontol Scand.2008;66(4),193-199.
8. Canger EM, Çelenk P, Yenísey M, Odyakmaz SZ. Amelogenesis Imperfecta, Hypoplastic Type Associated with Some Dental Abnormalities: A Case Report. Braz Dent J.2010;21(2):170-174.
9. Atar M, Körperich EJ. Systemic disorders and their influence on the development of dental hard tissues: a literature review. J Dent 2010;38(4):296-306.
10. Pousette Lundgren G, Wickström A, Hasselblad T, Dahllöf G. Amelogenesis Imperfecta and Early Restorative Crown Therapy: An Interview Study with Adolescents and Young Adults on Their Experiences. PLoS One. 2016;11(6).
11. Chen CF, Hu JC, Bresciani E, Peters MC, Estrella MR. Treatment considerations for patient with Amelogenesis Imperfecta: a review. Braz Dent Sci. 2013;16(4):7-18.
12. Nigam P, Singh VP, Prasad K, Tak J, Sinha A, Grewal P. Amelogenesis imperfecta - A review. J Adv Med Dent Sci Res 2014;2:83-90.
13. Ranganath V, Nichani AS, Soumya V. Amelogenesis imperfecta: A challenge to restoring esthetics and function. J Indian Soc Periodontol. 2010;14(3):195-7.
14. Singhal R, Pathak A, Goenka P. Amelogenesis Imperfecta with Anterior Open Bite: A Rare Case Report. Int J Clin Pediatr Dent. 2011;4(3):245-7.
15. Zupan AB, Sedano H, Scully C. Dento/Oro/ Craniofacial Anomalies and Genetics. Elsevier.2012:115-116.

Volume 2, Issue 2; March-April 2019; Page No. 85-92