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A Review Article:

Amelogenesis imperfecta; genes causes and treatment

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Abstract

Amelogenesis imperfecta (AI) is a group of disorder that causes disturbance in enamel formation resulting in development alterations of mineralization. Restoration of the dentition poses a great challenge when all the teeth are severely affected. The treatment opportunities for these patients may help in refining their lifestyle. The total treatment modality aims to relieve the pain and sensitivity, and preserving the tooth structure as much as possible. This article discusses both the structural and aesthetic rehabilitation of a patient with different types of AI.

Key words: *Amelogenesis imperfecta , Dental anomalies, Full mouth rehabilitation , Occlusal wear.*

1. Introduction :-

Amelogenesis imperfecta (AI) refer to a group of rare genetic disorder that is characterised by abnormal enamel formation caused by genetic mutations that alter the quality of enamel [1]. It may affect all or some teeth in the deciduous and /or permanent dentition [2]. X-linked , Autosomal dominant and recessive modes of inheritance have been documented [3]. According to Witkop's classification [4] (Table 1) , the defects may be classified as hypocalcified , hypomaturative and hypoplastic types .The hypocalcified varieties are characterised by alterations in enamel mineralization . The enamel appears discoloured and soft and can be easily removal . The hypomaturative types of AI are associated with abnormalities of the maturation stages of enamel formation, resulting in the enamel being opaque and chalky in appearance [4,5,6] . The hypoplastic types are characterised by a deficiency in the quantity of enamel, which may be expressed clinically as thin enamel or pits or grooves on the enamel surface [7] .So, clinically, all AI appears as loss of dental structure, yellow, grey o brown teeth and dental hypersensitivity. Gingival alterations, dental eruption alteration and taurodontism are also frequent.

This condition is quite complex, and may produce social , aesthetic and functional problems that are not easy to diagnose and treat. [8]

To determine the presence of AI, an accurate diagnosis with other enamel defects and verification of alteration symmetric pattern linked to genetic inheritance are mandatory [9] . The main sequel to patients with AI is represented by dental sensitivity and breakdown of hard tissues due to weak mechanical properties of affected teeth [10] . Still, there are marked impacts on children and adolescents as a result of AI , including aesthetics, function and psychological aspect. Thus, attention should be taken to multi approach treatment, aiming to determine the correct immediate and long- term planning follow up[11].

1.1. Definition :

AI represents a group of conditions , genomics in origin, which affect the structure and clinical appearance of the enamel of all or nearly all teeth in a more or less equal manner, and which may be associated with morphologic or biochemical changes elsewhere in the body [12] .

1.2. Etiology :

Genetic , febrile illness or vitamin deficiency, local infection or trauma, fluoride ingestion, Congenital syphilis , Birth or idiopathic factors [13] .

1.3. Pathogenesis :

Enamel is the highly mineralized structure in the body with 85% of its volume occupied by hydroxyapatite crystals. ———>

During the organogenesis, the enamel metamorphose from a soft, pliable tissue to its final form which is almost devoid of protein. ———>

The final composition of enamel is a reflection of its unique molecular and cellular activity that takes place during its genesis. ———> Amelogenesis Imperfecta [14]

1.4. Classification And General features :

1.4.1.Hypoplastic AI :

Hypoplastic AI subtypes are characterised by the defective formation of enamel which is the primary feature. The hypoplastic types can be characterised by enamel that is pitted , has grooves or furrows, has large areas of missing, or enamel that is very thin over the entire tooth crown. Quantitative defect are seen when the enamel does not form in normal thickness either due to local or general factors . Clinically, the crown size varies from small to normal and small teeth may lack proximal contacts . The colour varies from normal to opaque white- yellow brown. Enamel may be rough, smooth, pitted , grooved , locally hypoplastic or the complete tooth crown may appear with thin enamel [14, 15,16] .

1.4.2.Hypomaturation AI :

In this type , Qualitative defect of the enamel is seen where the enamel is not sufficiently mineralized . The teeth are appear normal morphologically at the time of eruption, but eventually chip away post eruptively , especially in the occlusal areas. Clinically, the color of teeth here varies from creamy opaque to marked yellow/brown. The surface of the teeth appear soft and rough leading to sensitivity due to dentinal exposure. Open bite malocclusion is a common feature. The enamel thickness is normal but often chips off and abrades away easily. Radiographically , there appear to be reduce differentiation between enamel and dentin which may be difficult to verify. Enamel has contrast similar to greater than dentin , unerupted crowns have normal morphology radiographically [16,13].

1.4.3.Hypocalcified AI:

Qualitative defect occurs when the enamel is insufficiently mineralized and soft . In comparison with hypomaturation type, the mineralization in this type is markedly reduced . Clinically, the crowns of the teeth in such cases appear to be

opaque white to yellow-brown, soft rough enamel surface, dental sensitivity and very poor aesthetics [16,17] .

Due to sever hypomineralization , there may be early loss of enamel. The thickness of enamel appear to be normal at eruption that often chips and but ,tend to abrade easily post eruptively . There may be delayed eruption of teeth. An anterior open bite of skeletal origin may be seen . Accumulation of a large amount of supragingival calculus is evident [16, 17] .

1.4.4.Hypomaturation- Hypoplastic with Taurodontism :

Clinically, the crown appears to be white/ yellow-brown mottled. The teeth appear smaller than normal and they lack proximal contacts . In these cases, the enamel thickness is drastically reduced. The crowns show pitting and tend to have hypomineralized areas. Radiographically, the enamel contrast is normal to slightly greater than dentin , and show large or bulbous pulp chambers which appear taurodontic [15,16]. In Figure 1 showed (A) Hypoplastic AI: dental enamel with thin, yellow-brown ditches. (B) Hypomaturation AI: It shows a mottling with snowflakes appearance.(C,D) hypocalcified AI: yellow-brown soft, friable dental enamel.

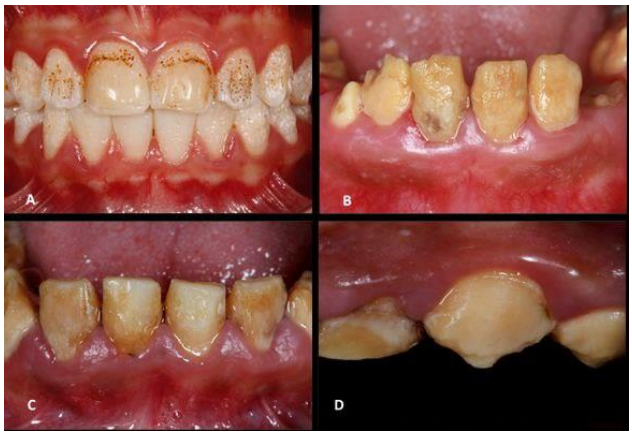

Figure 1 : Variations of dental phenotype in amelogenesis imperfecta

1.5.Genes Involved In Amelogenesis Imperfecta:

1.5.1. Genes involved in non-syndromic hypoplastic AI:

1.5.1.1. AMELX:

The AMELX gene (Amelogenin)has 7 exons and is located on the X chromosome in position Xp22.2. It encodes amelogenin , a protein rich in proline, glutamine, leucine and histidine, and constitute nearly 90% of the enamel organic matrix [18]. Amelogenin is phosphorylated , highly hydrophobic, and relatively basic , and self-assembles in spherical aggregate known as Amelogenin nanospheres . These nanospheres have an affinity for hydroxyapatite, controlling the direction of crystal in enamel and avoiding electrostatic interactions [19].

The AMELX transmission mode is giving by the X chromosome. Mutations in this gene cause X-linked AI [20].

In females it appears as a lionization phenomenon (random inactivation of one of the two X chromosome) responsible for an enamel with healthy and discoloured strips at the same time . In males there is a copy of AMELX as AMELY (Amelogenin) located in locus Yp11.2,responsible for encoding 10%of Amelogenin [21]. In the presence of AMELX gene can be a hypoplastic or hypomaturation AI in both deciduous and permanent dentition. Deletion and variant in the N-terminus cause hypomaturation AI with focal hypoplasia . On other hand, mutations in single peptide and toward the C-terminus portion are responsible for a hypoplastic AI [22].

Recently, Brookes pointed out that AI associated to the p. Tyr64H is mutation of the AMELX gene induce stress of the endoplasmic reticulum followed by apoptosis of ameloblasts . This cellular event causes an interruption of the secretory pathway of ameloblasts that form dentin , favoring the onset of AI . However, further studies are needed to accurately elucidate the dental alterations induced by mutations of the AMELX gene [23].

1.5.1.2. AMBN:

The AMBN gene (ameloblastin) consists of 13 exons and is located on chromosome 4, position 4p13.3. It encodes for ameloblastin, the second most abundant enamel matrix protein during Amelogenesis. AMBN is rich in glycine, leucine, and proline, is located in the Tomes process, but has also been detected in pre-odontoblasts[22].Ameloblastin is rapidly partitioned after the secretory stage , its fragments are incorporated in the prisms and works to avoid the fusion between prisms and the interprismatic substance.

AMBN participates in the differentiation and proliferation of ameloblasts, as well as in the extra cellular signalling that induces the differentiation of osteoblasts and cell adhesion [24,25,26]. Recently reported two mutations of the AMBN gene in patients with AI . The first is a deletion in exon 6, reducing the protein amino acids from 447 to 368, clinically, an aprismatic thin enamel was found. The second mutation produced the loss of exon7. This exon encodes a domain involved in the interaction of heparin and fibronectin , which are essential for the interaction of AMBN with epithelial cells [27,28,29].

1.5.1.3. ENAM:

The ENAM gene (enamelin) has 9 exons and is located on chromosome 4, position 4p13.3. It encodes for enamelin, a protein responsible for the nucleation and elongation of hydroxyapatite crystals. It expresses during the secretory stage mainly and degrades quickly from its terminal carboxyl end after its secretion by protases, producing enamelins of a lower molecular weight that are usually found in the prisms and interprismatic substance [30,31].

Recently reported two heterozygous mutations in the ENAM gene . Clinically, a hypoplastic AI with horizontal veins and loss of enamel substance was observed. Saymen et al [32] indicated two new heterozygous mutations of the ENAM gene. They also found that several individuals of the two analyzed families, despite having the same mutation as the affected individuals, presented incomplete penetrance.

1.5.1.4. LAMB3:

The LAMB3 gene (laminin beta-3) is composed of 23 exons and is located on chromosome 1, position 1q32.2. This gene encodes a protein known as laminin beta-3, a subunit of laminin 5. It acts as the transmembrane level (in the basal membrane) and participate in cell growth and adhesion . The enamel of patient with a mutation of the LAMB3 gene show speckles of varying extension “in a thimble form” on the surface of some or all teeth and in areas of vertical coloration [33].

1.5.1.5. LAMA3:

The LAMA3 gene (laminin alpha-3) consists of 75 exons and is located on chromosome 8, position 18q11.2. This gene encodes for laminin alpha-3. Mutations of this gene are related to syndromic and non- syndromic hypoplastic AI. Only two mutations of the LAMA3 gene related to non- syndromic AI have been reported, the first is located in exon 19 , while the second is located in exon 33. In both affected families, there was a hypoplastic AI in the absence of clinical dermatological signs [34].

1.5.1.6. ACPT:

The ACPT gene (acid phosphates , testicular) contain 11 exons and is located on chromosome 19, position 19q13.33. It encodes an enzyme capable of hydrolyzing orthophosphoric acid esters in acidic condition. By immunohistochemical analysis, it has been shown that ACPT is located in secretory ameloblasts, follicular cells, and osteoblasts [35]. Indeed, Choi et al [36] suggested that ACPT is capable of causing differentiation and mineralization of odontoblasts by supplying phosphate during dentin formation.

Seymen et al [35] identified 6 families with biallelic mutations of the ACPT gene. The homozygous mutations (c.713C>T; p.Ser238Leu, c.331C>T; P.Arg111Cys and c.226C>T; P.Arg76Cys) and two Compound heterozygous mutations (c.382G>C; P.Ala128Pro and 397G>A; P.Glu133Lys) were reported. In

addition, there were alterations in the size and lateral chains of the amino acids of the protein, which limit their accessibility to the catalytic nucleus and interfere with their homodimerization. Smith et al[37] describes two homozygous mutations (c.428C>T; P.T143M and c.746C>T; P. P249L) that were responsible for generalize hypoplastic AI. Both studies reported the role that ACPT can play in the secretory stage during amelogenesis, since the analysis showed the existence of a decrease in enamel (when it was present) but at the same time showed a well mineralized enamel.

1.5.2.Genes involved in non-syndromic hypocalcified AI:

1.5.2.1.FAM83H:

The FAM83H gene (family with sequence similarity 83, member H), composed of 5 exons, is located on chromosome 8, position 8q.24.3. It encodes for the intracellular FAM83H protein that participates in the differentiation of pre-ameloblasts in functional ameloblasts and in the mineralization process of enamel matrix. Its maximum expansion is found in secretory ameloblasts and the minimum in the ripening stage [38,39]. Its mode of transmission is autosomal dominant, and the resulting AI is hypocalcified, either localised or generalised. Clinically, deciduous and permanent teeth are affected and show mineralization defects characterized by a rough and porous dentin [40,41]. Xin et al[40] and Lee et al [42] showed that mutations of the FAM83H gene alter the location of the protein, presenting a higher concentration within the nucleus rather than in its cytoplasmic location. Kuga et al [43] showed that FAM83H regulates the organization of the cytoskeleton and maintains the formation of desmosomes. The authors suggest that, in the case of an AI resulting from an alteration of the FAM83H gene, there is a disorganisation of the cytoskeletal keratin with a subsequent alteration of the desmosomes at the ameloblastic level.

1.5.2.2. C4ORF26:

The C4ORF26 gene (Chromosome 4 open reading frame 26) is composed of 2 exons and is located on chromosome 4, position 4q21.1. It encodes a protein rich in proline of the extracellular matrix containing a single peptide with two highly conserved motifs and ten sites destined for phosphorylation. Based on the amino acid sequence, it has been estimated that C4ORF26 belongs to family of phosphoprotein and, according to Parry et al [44] this protein promotes the crystallisation of hydroxyapatite , supporting the growth of crystals after the phosphorylation of the C-terminus region .The mode of transmission is autosomal recessive and the phenotype corresponds to a hypocalcified. AI in deciduous and permanent teeth, showing a brownish yellow coloration, premature wear , and sensibility problems [45,44].

1.5.2.3. SLC24A4 :

The SLC24A4 gene (solute carrier 24 A4) is composed of 17 exons and is located on chromosome 14, position 14q32.12. It encodes a protein that functions as an ion exchanger (calcium/sodium/potassium dependent ion carrier). This protein has highly conserved hydrophobic regions (alpha-1 and alpha-2) that interact in the ions bonding located at the transmembrane level [46,47]. It expresses in the maturation ameloblasts and it has been suggested that it is responsible for the active transportation of Ca^{2+} ions from the ameloblasts to the enamel matrix during the maturation stage [48,49]. Missense mutations have been described in this gene affecting the alpha regions and the cytoplasmic domain, as well as multi-exonic deletions [47,49,50]. This gene expresses in a wide range of tissues, such as brain, aorta, lung and thymus.

1.5.2.4. ITGB6:

The ITGB6 gene (integrin B-6 chain) has 15 exons and is located on chromosome 2, position 2q24.2. It encodes for an integrin located in epithelial cells. This protein participates in the interactions between cells and MEC cells,

facilitating the interaction with the cytoskeleton [51].wang et al [52] suggest that ITGB6 is predominantly located in the ameloblasts of the maturation stage, and would play an essential role in fibronectin receptors and in the activation of matrix metalloproteinase 20 (MMP-20) or enamelysin. Clinically, it presents a hypoplastic AI with varying enamel compromise or a hypocalcified AI with loss of enamel substance and exogenous discoloration such as those described by Wang et al [52] and Poulter et al [53]. Its mode of transmission was autosomal recessive.

1.5.2.5. AMTN:

The AMTN gene (amelotin) contains 9 exons and is located on chromosome 4, position 4q13.3. It encodes a protein rich in proline, leucine, threonine and glutamine, secreted by ameloblasts in maturation stage during enamel formation. Barlette and Simmer [54] estimated that AMTN forms aggregates that mediate the bonding between maturation ameloblasts and mineralized enamel. An expression of AMTN in the junctional epithelium has also been reported. The phenotypic analysis showed a hypocalcified AI with the enamel presenting a weak mineral density and an altered structure in its entire extension[55,56].

1.5.3.Genes involved in non-syndromic hypomaturation AI:

1.5.3.1.MMP20:

The MMP20 gene (matrix metalloproteinase, enamelysin) has 10 exons and is located on chromosome 11, position 11q22.2. It encodes for enamelysin, a protein involved in cell motility and organic matrix degradation during the enamel maturation phase. The action of MMP20 directs the morphology of hydroxyapatite crystals and induces the increase in thickness of the hydroxyapatite crystals of dentin [57,58]. The MMP20 can act on the extracellular domains of cadherins that allow cell-cell interactions as part of the adherent joints in the movement of ameloblastic cells. This influence amelogenesis since ameloblasts should be arranged in a synchronised manner to form a normal ameloblastic structure. The inheritance mode is autosomal recessive and the phenotype is a pigmented

hypoplastic or hypomaturation AI. Homozygous (c.323A>G, c.954-2A>T, c.678T>A) and compound heterozygous mutations (c.567T>C, c.910G>A, c.126+6T>G, c.389C>T, c.954-2A>T and c.540T>A) have been reported as responsible for a porous, opaque, yellow enamel with sever wear and occasional mottling [59,60].

1.5.3.2. KLK4:

The KLK4 gene (Kallikrein 4) is composed of 5 coding exons and is located on chromosome 19, position 19q13.41. It encodes a serine protease that is expressed and secreted by ameloblasts in the maturation stage during amelogenesis. This serine protease participates in the nucleation and mineralization of enamel [61]. The MMP20 can activate the newly secreted KLK4 and in turn KLK4 can inactivate the MMP20, an event that explains the change in protein activity during the maturation stage. While contributing to the elimination of enamel proteins, KLK4 allows the growth of hydroxyapatite crystals [62].

1.5.3.3. WDR72:

The WDR72 gene (protein 72 with repeated WD) consists of 20 exons and is located on chromosome 15, position 15q21.3. It encodes a protein that acts at level of cell membranes during enamel mineralization. It expression is more intense in the maturation stage than in the secretory stage during dental development [63]. It mode of transmission is autosomal recessive, accompanied by a hypomaturation AI. The dentin shows a normal thickness but with premature wear and yellow-brown color. A delay in dental eruption and even short stature has been described in patients suffering from mutation of the WDR72 gene [63,64].

1.5.3.4. STIM1:

The STIM1 gene (stromal interaction molecule 1) consists of 12 exons and is located on chromosome 11, position 11p15.4. It encodes a transmembrane protein with calcium binding domains, located in the endoplasmic reticulum. This protein is a calcium sensor that allows the transfers of calcium ions from the endoplasmic

reticulum to the cell membrane. The STIM1 protein mediate the store-operated calcium entry (SOCE) , which is necessary for the normal functioning of ameloblasts. The expression of STIM1 has been highly detected in ameloblasts maturation, compared with ameloblasts secretion during dental formation [49]. Homozygous mutations with loss of function of the STIM1 gene cause combined immunodeficiency (CID) by STIM1 deficiency, a form of CID due to a dysfunction of calcium release-activated channels (CRAC). Patients with these mutations show hypocalcified AI leading to dentin loss [65].

1.5.3.5. GPR68:

The GPR68 gene (G protein-coupled receptor 68) has 1 exon and is located on chromosome 14, position 14q32.11. It encodes a proton-sensitive protein containing seven transmembrane helices, with histidine residues responsible for pH detection on the extracellular surface [66]. Parry et al [67] determined that GPR68 is expressed in the ameloblasts during all the amelogenesis stage.

Two homozygous mutations (c.667-668delAA; p.Lys223Glyfs and c.221T>C; p.Leu74Pro) in the only exon of the GPR68 gene produce a hypomaturation AI, characterised by an opaque enamel, anterior open bite , and a loss of dental substance in permanent teeth [68].

Table 1: Genes involved in non-syndromic amelogenesis imperfect:

	Gene (OMIM)	Position	Number of exons transmission	Encoded protein	AI type/associated syndrome
Hypoplastic AI	AMELX(amelogein) OMIM 300391	Xp22.2	7 linked to X	Amelogein (enamel matrix protein)	Hypoplastic or hypocalcified)
	AMBN(ameloblastin) OMIM 601259	4q13.3	13-AR	Ameloblastin (enamel matrix protein)	Hypoplastic
	ENAM(enamelin) OMIM 606585	4q13.3	9-AD/AR	Enamelin(enamel matrix protein	Hypoplastic
	LAMB3(laminin Beta-3) OMIM 150310	1q32.2	23-AD	Laminin beta-3	Hypoplastic

	Gene (OMIM)	Position	Number of exons transmission	Encoded protein	AI type/associated syndrome
	LAMA3(laminin alpha-3) OMIM 600805	18q11.2	75-AD	Laminin alpha-3	Hypoplastic
	ACPT(acid phosphates, testicular) OMIM 606362	19q13.33	11-AR	Testicular acid phosphatase	Hypoplastic
Hypocalcified AI	FAM83(family with sequence similarity 83, member H)OMIM 611927	8q24.3	5-AD	Intracellular protein- involved in ameloblastic differentiation	Hypocalcified
	C4ORF26(chromosome 4 open reading frame 26) OMIM 614829	4q21.1	2-AR	Extracellular matrix acid phosphoprotein	Hypocalcified
	SLC24A4(solutcarrier 24A4) OMIM 609840	14q32.12	17-AR	Ion exchanger	Hypocalcified
	ITGB6(integrin B-6chain) OMIM 147558	2q24.2	15-AR	Integrin of epithelial cells	Hypocalcified
	AMTN(amelotin) OMIM 610912	4q13.3	9-AD	Amelogein (enamel matrix protein)	Hypocalcified
Hypomaturation AI	MMP20(enamelysin) OMIM 604629	11q22.2	10-AR	Metalloproteinase	Hypomaturation
	KLK4(kalikrein4) OMIM 603767	19q13.41	6-AR	Serine protease	Hypomaturation
	WDR72(protein 72 with repeated WD) OMIM 612314	15q21.3	20-AR	Cytoplasmic protein- enamel mineralisation	Hypomaturation
	SITM1(stromal interaction molecule 1) OMIM 605921	11p15.4	12-AR	Transmembrane protein- calcium sensor	Hypomaturation
	GPR68(G protein- coupled receptor 68) OMIM 6015921	14q32.11	1-AR	Enamel matrix pH sensor	Hypomaturation

1.6. Syndrome Associated With AI :-

AI with taurodontism is found to be associated with Trichodentoosseous (TDO) syndrome. TDO syndrome is a AD condition characterized by splitting of the superficial layers of nails, kinky or tightly curled hair, bone sclerosis of long bones and skull base, zones of provisional calcification in the long bone, taurodontism , and enamel hypoplasia that occurs with Hypomaturation/hypo calcification defects [69].

2. Diagnostic principles behind AI:-

Accurate diagnosis requires a proper clinical history, clinical examination so that the presence of certain systemic disease that may show generalized enamel hypoplasia can be excluded, identification of mode of inheritance determined by family pedigrees chart and proper radiographic interpretation. Accurate diagnosis enables genetic counseling in an early phase, and precautionary steps can be taken as an early step to prevent further dental complications for the patient and even for upcoming siblings in the future. Historical confirmation can be done, but required extraction of the affected tooth, which is not a good idea in all cases except where prognosis of that tooth is poor.

3. Management :-

Management directed at three aspects of treatment including prevention, restoration, and esthetic. Most of the time patient report to the dentist when the dental complication like dental sensitivity or dental caries would have been started, so restoration should be undertaken as a first step [69-74].

Prevention aspects include dietary advice, regular use of fluoride mouthwashes, topical fluoride applications and oral hygiene instructions. Oral hygiene can be difficult for these patients due to the sensitivity while brushing so

warm water for teeth brushing can be advised. Along with prevention measures, long-term clinical follow up term is mandatory.[69-74]

Restoration aspects can be divided at the time of dentition, I.e., during primary dentition, direct

composite veneers for anterior teeth, GIC and stainless steel crowns for primary molars can be advised while in the mixed dentition stainless steel crowns, onlays for permanent molars , composite or GICs for primary and permanent teeth and for permanent incisors, direct or indirect composite veneers should be advised.[69-74]

The esthetic treatment modality also decided based on dentition, I.e., minimal intervention GIC restoration should be advised in the primary dentition, direct and indirect composite resin veneers in the mixed dentition while porcelain veneers, full crowns, extraction of excessive defected teeth followed by fixed, or removable prosthesis should be advocated in permanent dentition based on number of teeth affected, patients age, and economical status.[69-74]

Still no standard formula, current protocol, or guideline for successful treatment the accurate treatment should be planned based on type of AI , severity and oral health habits of the patient. Except for mildly affected teeth, restoration with amalgam are usually unsuccessful due to fracture of the weak enamel margins. Adherent materials such as glass ionomer cements and composite resins are better retained compared to amalgam restoration. However, in cases of hypocalcified AI where the enamel is very weak and bonding of the restoration is questionable, full coverage is required. In the primary and early mixed dentition, stainless steel crowns are effective restoration.[19-24] The most effective methods for dentinal sensitivity is full coronal coverage using stainless steel crowns on the posterior teeth, especially in mixed dentition and primary dentition.[69-74]

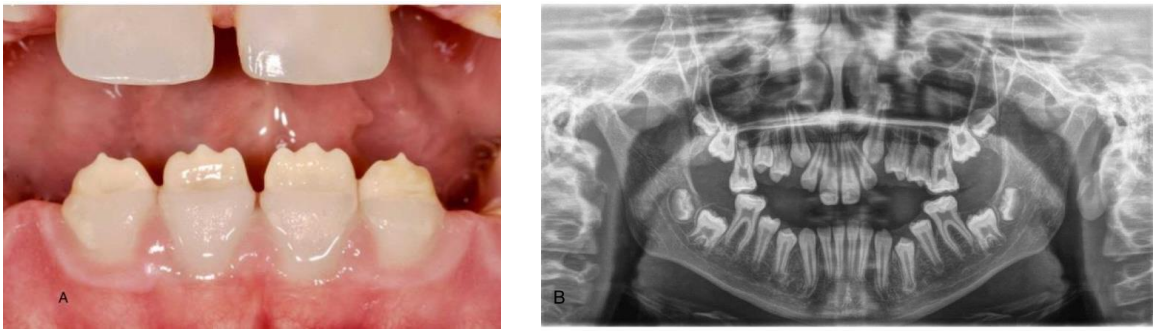

Figure 2: Preoperative view of lower incisors affected by AI type 1(A), preoperative panoramic radiographic view(B).

In constructing steel crowns, a conservative technique of tooth separation using separating elastics prior to the insertion of the crowns is recommended. Glass ionomer cements are likely to be better luting agents for the crowns compared with zinc phosphate if there are large areas of exposed dentin. Anterior open bite can be treated with different treatment modalities ranging from orthodontic banding or orthoganthic surgery. Orthodontic treatment can be performed only after once all restoration treatment is finished[69-74].

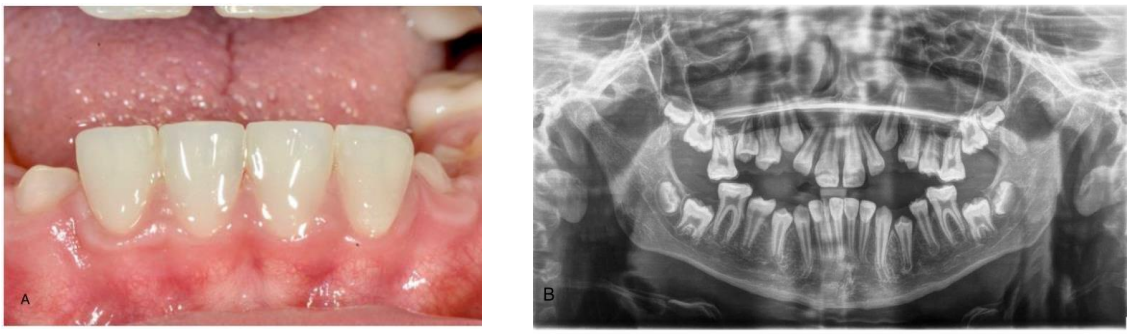

Figure 3: postoperative view of Edelweiss Veneers (A), postoperative panoramic radiographic view (B).

4. Result:-

Nineteen eligible articles were included in this study. Management includes removal of surface stains, reducing sensitivity, maintaining vertical dimension of occlusion, and the esthetic with adhesive techniques/ over denture/ porcelain- fused to metal crowns / fixed partial dentures / full porcelain crowns/ inlay / onlay restorations are used for better esthetics of the patients. Preventive aspects in the

primary and mixed dentition include dietary advice, fluoride supplements and oral hygiene instruction. Restorative to prosthodontics density must be done in order to maintain oral function and growth preventing tooth loss and allowing oral hygiene maintenance. The first consultation must be as early as possible. Treatment alternatives deal with minimal invasive dentistry with the objective of maintaining tooth vitality as long as possible.

5. Conclusion:-

AI is an inherited disorder affecting enamel. This entity more often causes psychological stress in patients due to poor esthetic. A proper history, clinical evaluation, genetic mapping, radiological interpretation followed by best suitable treatment are the necessary steps to make them smile. Dental practitioners should be aware of all possible treatment for AI as one treatment which is successful in one type, may fail in others.

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